Circulation Journal
Official Journal of the Japanese Circulation Society
http://www.j-circ.or.jp

Study of the Association Between an Anomalous Superior Vena Cava and Horseshoe Kidney

Tamaki Ichikawa, MD, PhD; Tatsuya Sekiguchi, MD; Shuichi Kawada, MD; Jun Koizumi, MD, PhD; Jun Endo, MD, PhD; Yuri Yamada, MD; Chihiro Ito, MD; Makoto Sugiyama; Toshiro Terachi, MD, PhD; Yukio Usui, MD, PhD; Kojun Torigoe, MD, PhD; Yutaka Imai, MD, PhD

Background: The incidence of inferior vena cava anomalies in patients with horseshoe kidney is higher than that reported in the general population. As far as we know, no studies have reported the incidence and variations of superior vena cava (SVC) anomalies using multidetector-row computed tomography (MDCT) in patients with horseshoe kidney.

Methods and Results: Using MDCT, 71 patients with a horseshoe kidney (group A: 45 males, 26 females; mean age, 60.1±10.2 years) and 2,292 patients without a horseshoe kidney (group B: 1,385 males, 907 females; mean age, 61.1±13.5 years) were retrospectively evaluated for the incidence and variations of SVC anomalies, and the incidence of an anomalous SVC was compared between groups. An anomalous SVC was identified in 3 group A patients (4.2%) (double SVC, n=2; persistent left SVC without a right SVC, n=1) and 5 group B patients (0.22%) (double SVC, n=3; persistent left SVC without a right SVC, n=2). MDCT revealed a significantly higher incidence of anomalous SVC in patients with a horseshoe kidney than in those without a horseshoe kidney (P<0.001).

Conclusions: Patients with horseshoe kidney frequently have an anomalous SVC. Although the incidence of horseshoe kidney is related in some way to that of an anomalous SVC, the reasons for their coexistence remain unclear. (*Circ J* 2012; **76:** 1253–1258)

Key Words: Computed tomography; Horseshoe kidney; Superior vena cava

Horseshoe kidney is a well-known congenital anomaly involving the upper urinary tract. It occurs in 1 in 400–800 individuals and exhibits male predominance.[1,2] Horseshoe kidney is characterized by a fusion of the lower poles of both kidneys by parenchymal tissue, which is also known as the isthmus. This condition is rarely associated with anomalous inferior vena cava (IVC).[3–7] The incidence of IVC anomalies detected by multidetector-row computed tomography (MDCT) in patients with a horseshoe kidney is reportedly 5.7%, which is higher than that reported in the general population without a horseshoe kidney.[7] An anomalous superior vena cava (SVC) is also rarely associated with horseshoe kidney.[8,9] The development of the renal parenchyma and its drainage into the IVC occur simultaneously during gestational weeks 4–10, and the IVC and SVC are formed from cardinal veins around the same gestational period.[6,10] To our knowledge, an original article regarding anomalous SVC associated with horseshoe kidney has not been reported to date. In the present study, using MDCT, we evaluated the incidence and variations of SVC anomalies in 2 groups of patients: those with a horseshoe kidney and those without a horseshoe kidney. We then compared the results of both groups.

Methods

Informed consent was not required because this was a retrospective study approved by the institutional review board at Tokai University Hospital. The study involved 2 groups: group A comprised 71 patients with a horseshoe kidney (45 males, 26 females; mean age, 60.1±10.2 years) and group B comprised 2,292 patients without a horseshoe kidney (1,385 males, 907 females; mean age, 61.1±13.5 years). All patients in group A were diagnosed as having horseshoe kidney between January 2006 and November 2011. During this period, all these patients had undergone CT examination of the region between the chest and abdomen: 37 for the evaluation of urological symptoms and 34 for the evaluation of non-urological symptoms. Horseshoe kidney was discovered incidentally dur-

Received August 9, 2011; revised manuscript received December 7, 2011; accepted January 6, 2012; released online February 17, 2012
Time for primary review: 57 days

Department of Radiology (T.I., T.S., S.K., J.K., J.E., Y.Y., C.I., Y.I.), Department of Radiological Technology (M.S.), Department of Urology (T.T., Y.U.), and Department of Anatomy (K.T.), Tokai University School of Medicine, Isehara, Japan
Mailing address: Tamaki Ichikawa, MD, PhD, Department of Radiology, Tokai University School of Medicine, 143 Shimokasuya, Isehara 259-1193, Japan. E-mail: tamaki-i@mars.sannet.ne.jp

ISSN-1346-9843 doi:10.1253/circj.CJ-11-0874

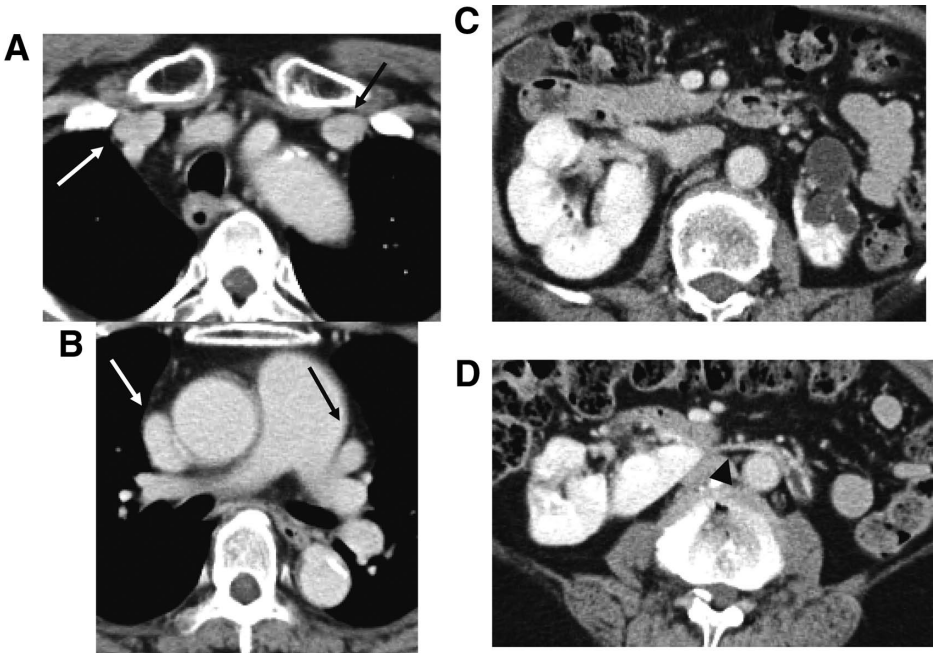

Figure 1. A 68-year-old female with a double superior vena cava (SVC) associated with a horseshoe kidney. (**A**,**B**) Chest computed tomography (CT) images with contrast enhancement reveal a right SVC (white arrows) and a left SVC (black arrow) with no bridging brachiocephalic vein. (**C**,**D**) Abdominal CT images with contrast enhancement reveal atrophy of the left side of the kidney with multiple cysts, hydronephrosis and a thin line of fusion between the lower poles (arrowhead).

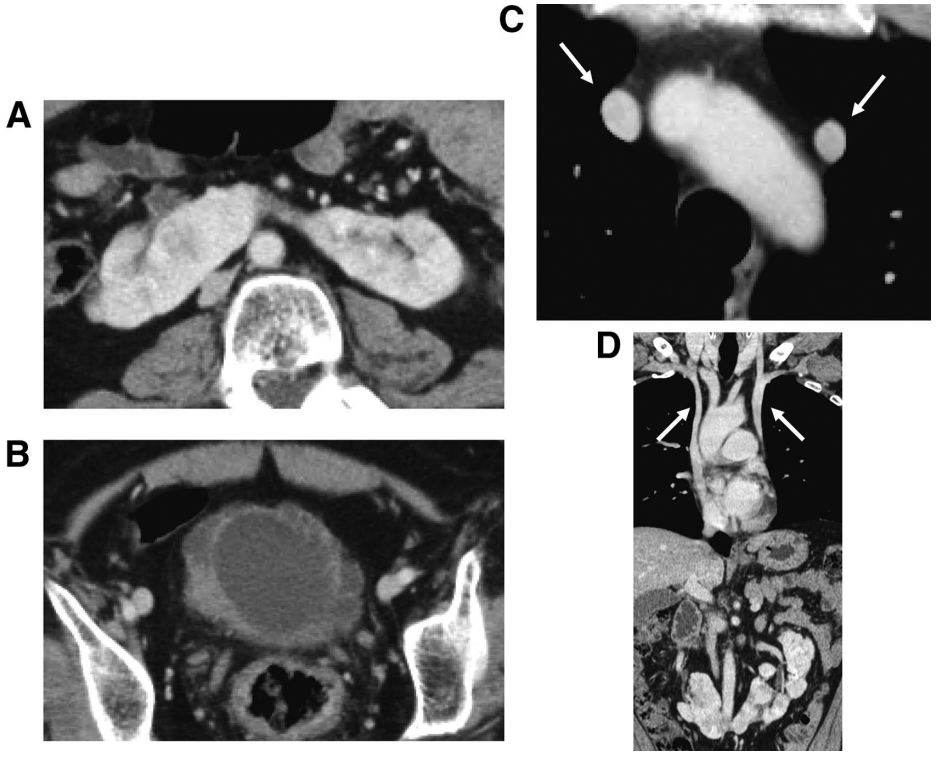

Figure 2. A 46-year-old male with a double superior vena cava (SVC) associated with a horseshoe kidney and spina bifida. (**A**,**B**) Abdominal computed tomography (CT) images with contrast enhancement show horseshoe kidney and a trabeculated bladder. (**C**,**D**) Chest CT images show a double SVC (arrows) with no bridging brachiocephalic vein.

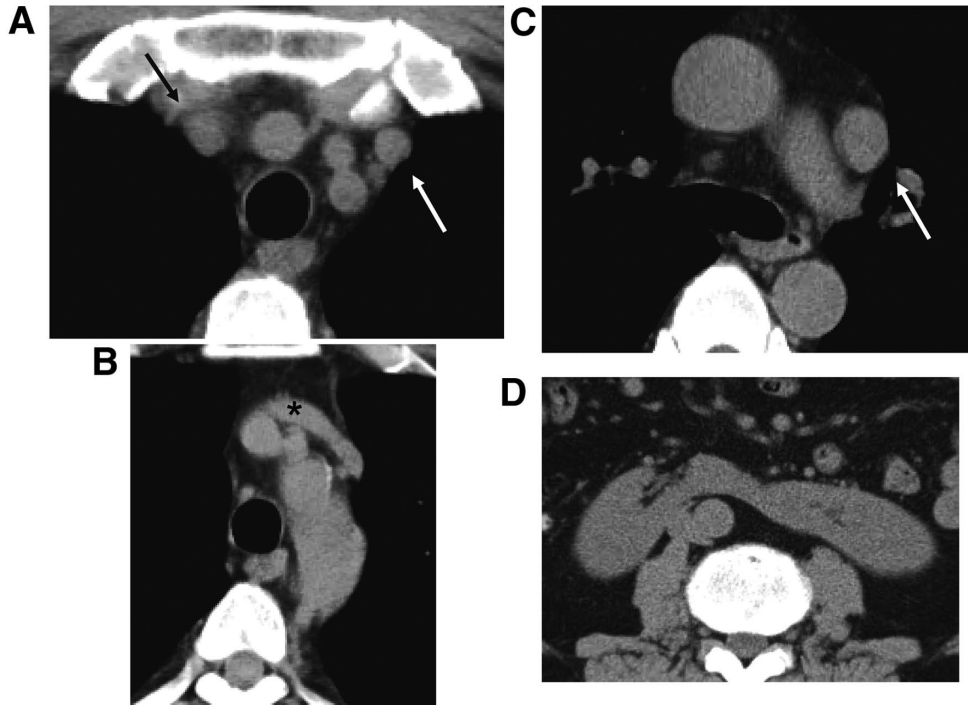

Figure 3. A 67-year-old male with a persistent left superior vena cava (SVC) and an absent right SVC associated with a horseshoe kidney. (**A**–**C**) Chest computed tomography (CT) images without contrast enhancement reveal a persistent left SVC (white arrow) and 2 bridging brachiocephalic veins (black arrow, ⋆). (**D**) Abdominal CT shows fusion of the lower poles of the kidneys.

Table. Variations in ASVC and Other Associated Diseases in Patients With and Without a Horseshoe Kidney

Case no.	Group	Age (years)	Sex	CE-CT	Type of ASVC	Urinary disease
1	A	67	M	+	II	–
2	A	68	F	–	IIIb	Hydronephrosis and atrophy of left side of kidney, renal cysts
3	A	46	M	–	IIIb	Membranous nephropathy, neurogenic bladder
4	B	84	M	–	II	–
5	B	67	M	–	IIIa	Renal cyst
6	B	58	F	+	IIIb	–
7	B	71	M	–	IIIb	–
8	B	65	M	+	II	Renal cyst

ASVC, anomalous superior vena cava; CE-CT, contrast-enhanced computed tomography.

ing these examinations. In group B, 2,292 patients had undergone routine CT examination of the region between the chest and abdomen between January and February 2011.

We used 2 64-slice MDCT scanners with 0.5- and 0.6-mm slice thickness (Aquilion; Toshiba, Tokyo, Japan and Somatom Cardiac Sensation 64; Siemens, Forchheim, Germany) for the MDCT studies. Other parameters included 120 kVp, 125 mA, and 0.5-s rotation time. Contrast material was used in 36 group A patients and 1,507 group B patients, who were intravenously administered 100–150 ml (Omnipaque 300; Daiichi-Sankyo, Tokyo, Japan) at the rate of 2–3 ml/s, with a 120-s scanning delay in some patients. Two radiologists with more than 12 years' experience in CT image interpretation reviewed the axial CT images with a 5-mm reconstruction interval on a picture archiving and communication system workstation. Consensus was achieved through discussion. Two reviewers recorded the presence and variations of SVC anomalies, which were classified as: type I, normal precardial venous arrangement; type II, single persistent left SVC with an absent right SVC and a bridging brachiocephalic vein; and type III, double SVC, either connected by an anastomosis (type IIIa) or unconnected (type IIIb).[11] We compared the incidence of anomalous SVC between groups using the chi-square test. In both groups, we assessed the association of an anomalous SVC with congenital heart disease and/or other clinical characteristics of the groups were compared by unpaired t-test. Statistical significance was set at $P<0.05$.

Results

An anomalous SVC was identified in 3 group A patients (4.2%) and 5 group B patients (0.22%); the anomalies included

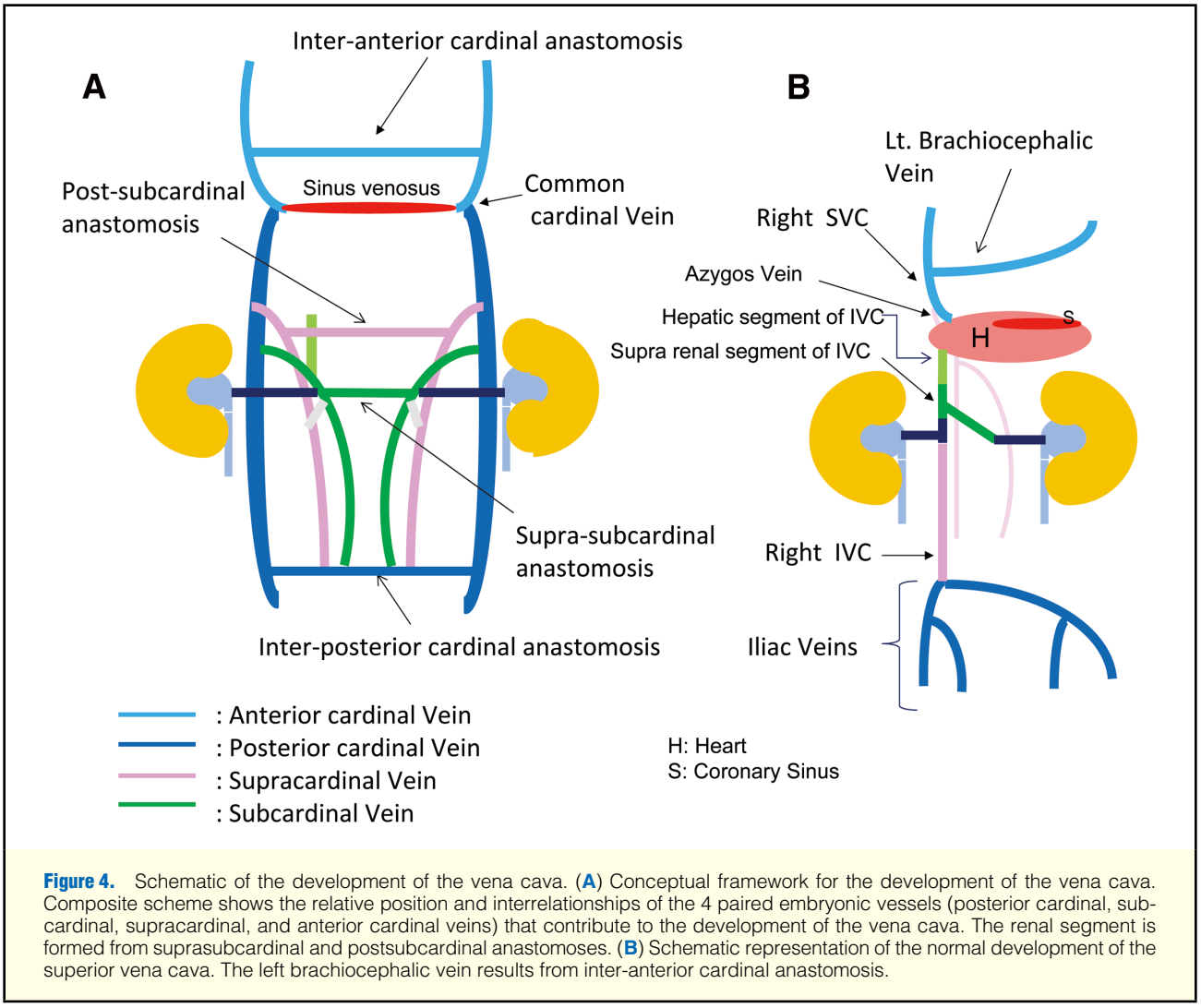

Figure 4. Schematic of the development of the vena cava. (**A**) Conceptual framework for the development of the vena cava. Composite scheme shows the relative position and interrelationships of the 4 paired embryonic vessels (posterior cardinal, subcardinal, supracardinal, and anterior cardinal veins) that contribute to the development of the vena cava. The renal segment is formed from suprasubcardinal and postsubcardinal anastomoses. (**B**) Schematic representation of the normal development of the superior vena cava. The left brachiocephalic vein results from inter-anterior cardinal anastomosis.

a double SVC (type IIIb, n=2) and a persistent left SVC (type II, n=1) (**Figures 1–3**) in group A and a double SVC (type IIIa, n=1; type IIIb, n=2) and a persistent left SVC (type II, n=2) in group B. MDCT indicated that the incidence of an anomalous SVC was significantly higher in patients with a horseshoe kidney than in those without a horseshoe kidney (P<0.001).

The various SVC anomalies, as well as other associated diseases, in each patient in both groups are listed in **Table**. One patient (case 3) with spina bifida and membranous nephropathy had required self-catheterization since childhood and had a neurogenic bladder (**Figure 2**). Two other patients with an anomalous SVC were incidentally found to have a horseshoe kidney during routine CT examination. However, 1 patient (case 2) had atrophy of the left side of the kidney with multiple cysts. An anomalous SVC was not associated with congenital heart disease in either group. No significant differences in the demographic data of the 2 groups were identified by unpaired t-test.

Discussion

Horseshoe kidney is the most common anomaly of the upper urinary tract,[1,2] characterized by fusion of the lower poles of both kidneys. This happens if the 2 metanephric blastemas come into contact, mainly at the lower pole, during migration from the sacral region. The bridge of parenchymal tissue connecting the lower poles is called the isthmus.[12] Typically, anterior displacement of the renal pelvis and ureter is observed in such cases. In addition, the ureters descend anterior to the isthmus and have a tendency to develop kinks, which often results in hydronephrosis.[12]

A wide variety of associated genitourinary and non-genitourinary anomalies coexist with this condition.[13,14] Association of horseshoe kidney with IVC anomalies, including double IVC, left IVC, and preisthmic IVC, is less frequently observed.[3–7] Retrocaval ureter is associated with preisthmic IVC, an anomaly that is the direct cause of hydronephrosis and ureteropelvic junction obstruction.[3] Because embryogenesis of the renal parenchyma and its venous drainage in the IVC occur simultaneously during gestational weeks 4–10, it is plausible that horseshoe kidneys and anomalous IVCs are consequences of a shared disturbed signal that occurs during the development of these retroperitoneal structures.[6]

Glodry et al reported a case of a persistent left SVC with a fusion anomaly of the kidney, but it is unclear if the anomaly was horseshoe kidney or crossed fused ectopia.[13] Two other cases of an anomalous SVC with horseshoe kidney have been reported.[8,9] One was a case of a persistent left SVC with horse-

shoe kidney,[8] and the other was a case of Prune belly syndrome in a stillborn fetus with a ring X chromosome lacking the X inactive-specific transcript.[9] The fetus had a persistent left SVC, horseshoe kidney, a single umbilical artery, limb deficiencies, cardiac hypertrophy, and axial skeletal abnormalities.[8] Both cases were autopsy reports.[8,9] Konishi and Kikuchi reported that coexistence of a persistent left SVC and horseshoe kidney in the same body is extremely rare.[8]

Horseshoe kidney is associated with IVC anomalies because their development occurs around the same gestational period.[6] The SVC and IVC differentiate around gestational weeks 4–10. The SVC derives from the anterior and common cardinal veins, whereas the IVC derives from the supra- and subcardinal veins (**Figure 4**). The coexistence of an anomalous SVC and an IVC in patients with normal kidneys has not been reported.

Horseshoe kidney and an anomalous SVC are both associated with a high incidence of cardiovascular malformations.[10,15–19] Therefore, it is reasonable to surmise that the incidence of horseshoe kidney is related in some way to that of an anomalous SVC. It is conceivable that SVC anomalies and horseshoe kidney may share the same etiology.

The most common congenital SVC anomaly is a broad and persistent left SVC (persistent left SVC, including double SVC). The incidence of this anomaly in the general population is 0.1–0.5%.[10,11,18–25] In our study, the incidence of an anomalous SVC in patients without a horseshoe kidney was 0.22%, a result similar to that of previous studies.[10,11,18–25] A persistent left SVC represents a failure of obliteration of the left anterior cardinal vein in the early stages of embryological development (**Figure 4**).[10] A broad and persistent left SVC is more common in patients with congenital heart disease (4–11%).[10,18,19] However, no congenital heart disease was present in any of the patients with an anomalous SVC in our study.

A persistent left SVC with an absent right SVC in normal situs is exceedingly rare, accounting for only 0.07–0.13% of congenital cardiovascular malformations.[25] A persistent left SVC and an absent right SVC were identified in 3 patients in our study, 1 of whom also had a horseshoe kidney. Incidence of a persistent left SVC with an absent right SVC associated with horseshoe kidney was therefore 1.5%. The incidence of this factor and the overall incidence of an anomalous SVC were higher in group A than in group B in our study. A persistent left SVC is often asymptomatic and hemodynamically insignificant. In most cases, it drains into the right atrium via the coronary sinus.[10] A bridging brachiocephalic vein is observed in approximately 30% of double SVC cases.[22] Central venous catheter (CVC) misplacement in cases of persistent left SVC increases the occurrence of mechanical and thrombotic complications.[11,22,23] CVC placement or access through the persistent left SVC may pose several serious problems, such as hypotension, angina, arrhythmias, and cardiac arrest.[11,20,23] In cases of persistent left SVC with an absent right SVC, pacemaker implantation is extremely difficult, even impossible.

More than half of the patients with horseshoe kidney have other urogenital, gastrointestinal, cardiopulmonary, skeletal, or chromosomal anomalies.[13,14] Sepsis secondary to pyelonephritis is the most severe complication in patients with a horseshoe kidney. Horseshoe kidney is associated with a higher probability of traumatic processes because of its location, anatomical factors, and the variability of vascularization.[26,27] There is an increased risk of rupture by compression of the isthmus against the spinal column or by fracture of the isthmus.[26] Therefore, careful surgical management and CVC placement are more critical in patients with a horseshoe kidney than in those without a horseshoe kidney. An anomalous SVC is a risk factor not only for central CVC insertion but also for pacemaker implantation.[11,20–23,28,29] Detection and knowledge of SVC anomalies in patients with a horseshoe kidney are crucial to reduce serious complications during CVC procedures, intracardiac electrode placement, and cardiopulmonary bypass.

Study Limitations

First, almost all patients were adults, and the sample of patients with a horseshoe kidney was small. Second, the presence of an anomalous SVC was evaluated by MDCT; surgical confirmation of venous anomalies was not performed. A large study involving autopsies on cadavers of all age groups is necessary to determine the true incidence of an anomalous SVC in patients with a horseshoe kidney.

Conclusions

We evaluated the incidence of an anomalous SVC in patients with a horseshoe kidney using MDCT. The incidence of an anomalous SVC was higher in patients with a horseshoe kidney than in those without a horseshoe kidney. The incidence of horseshoe kidney is related in some way to that of an anomalous SVC, but the reasons for their coexistence remain unclear.

References

1. Yoshinaga K, Kodama K, Tanii I, Toshimori K. Morphological study of a horseshoe kidney with special reference to the vascular system. *Anat Sci Int* 2002; **77:** 134–139.
2. Faggioli GL, Freynir A, Pilato A, Ferrii M. Curti T, Paragona O, et al. Renal anomalies in aortic surgery: Contemporary results. *Surgery* 2003; **133:** 641–646.
3. Youssif M. Horseshoe kidney with retrocaval ureter. *Eur Urol* 1985; **11:** 61–62.
4. Kehagias DT, Goulliamos AD, Vlahos LJ. Horseshoe kidney associated with anomalous inferior vena cava. *Eur Radiol* 1999; **9:** 935–936.
5. Smith T, Frost A. Anomalous inferior vena cava associated with horseshoe kidneys. *Clin Imaging* 1996; **20:** 276–278.
6. Radermecker MA, Van Damme H, Kerzmann A. Association of abdominal aortic aneurysm, horseshoe kidney, and left-sided inferior vena cava: Report of two cases. *Vasc Surg* 2008; **47:** 645–648.
7. Ichikawa T, Kawada S, Koizumi J, Endo J, Iino M, Terachi T, et al. Major venous anomalies are frequently associated with horseshoe kidneys: Value of multidetector computed tomography. *Circ J* 2011; **75:** 2872–2877.
8. Konishi M, Kikuchi M. A case of persistent left superior vena cava with a horseshoe kidney. *Kaibogaku Zasshi* 1991; **66:** 525–536 (in Japanese).
9. Guillen DR, Lowichik A, Schneider NR, Cohen DS, Garcia S, Zinn AR. Prune-Belly syndrome and other anomalies in a stillborn fetus with a ring X Chromosome lacking XIST. *Am J Med Genet* 1996; **70:** 32–36.
10. Minniti S, Visentini S, Procacci C. Congenital anomalies of the venae cavae: Embryological origin, imaging features and report of three new variants. *Eur Radiol* 2002; **12:** 2040–2055.
11. Schummer W, Schummer C, Frober R. Persistent left superior vena cava and central venous catheter position: Clinical impact illustrated by four cases. *Surg Radiol Anat* 2003; **25:** 315–321.
12. Oktem H, Gozil R, Calgunar E, Bahcelioglu M, Mutlu S, Kurkcuglu A, et al. Morphometric study of a horseshoe kidney. *Med Princ Pract* 2008; **17:** 80–83.
13. Glodny B, Petersen J, Hofmann KJ, Schenk C, Herwig R, Trieb T, et al. Kidney fusion anomalies revisited: Clinical and radiological analysis of 209 cases of crossed fused ectopia and horseshoe kidney. *BJU Int* 2009; **103:** 224–235.
14. Grainger R, Lane V, Murphy DM. Horseshoe kidney: A review of the presentation, associated congenital anomalies and complications in 73 patients. *Ir Med J* 1983; **76:** 315–317.
15. Greenwood RD, Rosenthal A, Nadas AS. Cardiovascular malformation associated with congenital anomalies of the urinary system: Observations in a series of 453 infants and children with urinary system

malformation. *Clin Pediatr* 1976; **15:** 1101–1105.
16. Voisin M, Djernit A, Morin D, Grolleau R, Dumas R, Jean R. Congenital heart disease and urinary tract malformations. *Arch Mal Coeur* 1988; **81:** 703–707.
17. Boatman DL, Kolln CP, Flocks RH. Congenital anomalies associated with horseshoe kidney. *J Urol* 1972; **107:** 205–207.
18. Nsah EN, Moore W, Hutchins GM. Pathogenesis of persistent left superior vena cava with a coronary sinus connection. *Pediatr Pathol* 1991; **11:** 261–269.
19. Buirski G, Jordan SC, Joffe HS, Wilde P. Superior vena cava abnormalities: Their occurrence rate, associated cardiac abnormalities and angiographic classification in a paediatric population with congenital heart disease. *Clin Radiol* 1986; **37:** 131–137.
20. Bartram U, Van Praagh S, Levine JC, Hines M, Bensky AS, Van Praagh R. Absent right superior vena cava in visceroatrial situs solitus. *Am J Cardiol* 1997; **80:** 175–183.
21. Peltier J, Destrieux C, Desme J, Renard C, Remond A, Velut S. The persistent left superior vena cava: Anatomical study, pathogenesis and clinical considerations. *Surg Radiol Anat* 2006; **28:** 206–210.
22. Ratliff HL, Yousufuddin M, Lieving WR, Watson BE, Malas A, Rosencrance G, et al. Persistent left superior vena cava: Case reports and clinical implications. *Int J Cardiol* 2006; **113:** 242–246.
23. Bhatti S, Haheem A, Ahmad U, Malik M, Kosolcharoen P, Chang SM. Persistent left superior vena cava (PLSVC) with anomalous left hepatic vein drainage into the right atrium: Role of imaging and clinical relevance. *Vasc Med* 2007; **12:** 319–324.
24. Benson RE, Songrug T. CT appearance of persistent left superior vena cava, anomalous right superior pulmonary venous return into the right-sided superior vena cava and a sinus venous-type atrial septal defect. *Br J Radiol* 2009; **82:** e235–e239.
25. Lenox CC, Zuberbuhler JR, Park SC, Neches WH, Mathews RA, Fricker FJ, et al. Absent right superior vena cava with persistent left superior vena cava: Implications and management. *Am J Cardiol* 1980; **45:** 117–122.
26. Murphy JT, Borman KR, Davidson I. Renal autotransplantation after horseshoe kidney injury: A case report and literature review. *J Trauma* 1996; **40:** 840–844.
27. Singer A, Simmons MZ, Maldjian PD. Spectrum of congenital renal anomalies presenting in adulthood. *Clin Imaging* 2003; **32:** 183–191.
28. Chen PS, Joung B, Shinohara T, Das M, Lin SF. The initiation of the heart beat. *Circ J* 2010; **74:** 221–225.
29. Lee YK, Sim JY, Seo JW, Choi IC, Hahm KD, Choi JW. Optimal placement of a superior vena cava cannula in minimally invasive robot assisted cardiac surgery. *Circ J* 2010; **74:** 264–268.