

Assessment of Renal Shape of Horseshoe Kidney with Multidetector Row CT in Adult Patients: Relationship between Urolithiasis and Renal Isthmus

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Objective: The aim of this study was to evaluate the relationship between urolithiasis and characteristics of renal shape in adult patients with horseshoe kidney (HSK) diagnosed on multidetector row computed tomography (MDCT).

Methods: We evaluated 36 patients with HSK and urolithiasis (Group A) and 70 patients with HSK without urolithiasis (Group B) whose disease was diagnosed on non-contrast MDCT. Two radiologists measured minimum width of the renal isthmus and maximum length of the renal pelvis and evaluated coexisting neoplastic diseases on axial computed tomographic (CT) images with 5-mm reconstruction, and we compared those measurements between the Groups A and B.

Results: The overall mean maximum length of the renal pelvis, 12.7 ± 9.2 mm, did not differ significantly between the 2 groups. Minimum isthmus width was larger in patients with HSK and urolithiasis (11.0 ± 5.6 mm), than those without urolithiasis (9.5 ± 5.1 mm). No patient in either groups had a urological renal tumor.

Conclusions: Patients of HSK might have tendency of a high incidence of stone formation. Because urolithiasis is a risk factor for tumors of the renal pelvis, monitoring of patients with HSK requires careful attention to isthmus width on CT images.

Key words: computed tomography, horseshoe kidney, isthmus, urolithiasis

INTRODUCTION

Horseshoe kidney (HSK) is the most common fusion anomaly, with incidence of one in 400 to 666 reported based on autopsy and radiologic series [1, 2]. HSKs are generally linked at the lower poles by an isthmus of parenchyma [1, 2] and are associated with a wide variety of coexisting anomalies [2–9]. Most adult patients with HSK are asymptomatic, and the anomaly is noticed incidentally on radiologic examination, such as ultrasound or computed tomography (CT) [2–4]. However, anatomical abnormalities, such as high insertion of the ureter or its anterior course over the isthmus, predispose the patient to impaired drainage of the collecting system, urinary stasis, and increased incidence of ureteropelvic junction (UPJ) obstruction and stone formation [1, 5, 6].

Urolithiasis is the most common complication of HSK, with reported incidence of 20 to 60% [5, 9], and is believed to occur most commonly secondary to urinary tract infection and stasis related to the abnormal anatomy of the UPJ and aberrant ureteric course, such as high insertion into the renal pelvis [4–6, 10]. HSK is difficult to diagnose by sonography, especially if the isthmus is not seen, but HSK is usually identified easily on CT [11]. Renal shape including collecting system depends on isthmus width [7]. We evaluated whether urolithiasis was related to characteristics of

renal shape, especially minimum width of the isthmus, of the HSK on multidetector row CT (MDCT) in adult patients.

PATIENTS AND METHODS

Informed consent was not required because this was a retrospective study approved by our institutional review board. Between January 2006 and December 2012, 131 adult patients were diagnosed with HSK using MDCT. We selected 106 patients who were performed non-contrast CT examinations of the region between the abdomen and pelvis and divided these patients into 36 patients (24 men, 12 women; aged 27 to 90 years, mean age 64.2 ± 14.8 years) with urolithiasis (Group A) and 70 patients (40 men, 30 women; aged 20 to 90 years, mean age 62.9 ± 17.3 years) without urolithiasis. Group A included 28 patients with renal stones, 7 patients with ureteral stones, and 2 patients with bladder stones; 1 patients with urolithiasis had both ureter and bladder stones. Contrast-enhanced CT was performed in 66 patients (Group A, 21; Group B, 45), computed tomographic (CT) angiography in 12 (Group A, one; Group B, 11), dynamic study of the upper abdomen in 18 (Group A, 8; Group B, 10), and CT urography in 11 (Group A, 5; Group B, 6).

MDCT was performed using 8- to 128-row MDCT scanners of 0.6- to one-mm slice thickness (LightSpeed, GE, Milwaukee, WI, USA; Somatom®



Fig. 1 The narrowest part of the connection between the halves of the kidney on axial image was considered the renal isthmus (arrow).

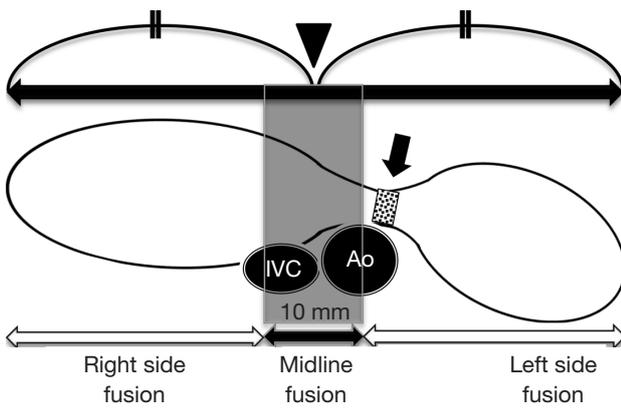


Fig. 2 Divide fusion site of the isthmus.
Isthmus: arrow, Center of the HSK: arrowhead
Midline fusion: less than 5mm on both sides from the center of the HSK
On this case, the isthmus is divided into the left side fusion.
IVC: inferior vena cava
Ao: aorta

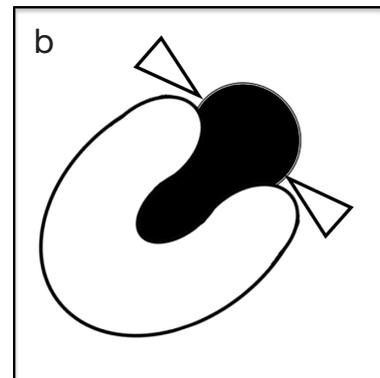
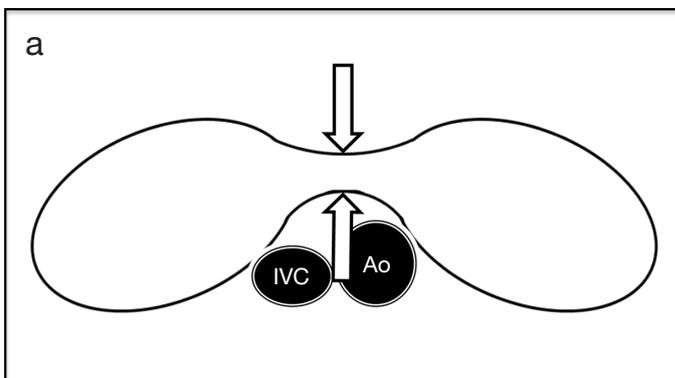


Fig. 3 Method of measurement
a. Minimum width of the renal isthmus (allows)
b. Maximum length of the renal pelvis (allowheads)
IVC: inferior vena cava
Ao: aorta

Sensation Cardiac 64, Definition, Definition FLASH, Siemens, Forcheim, Germany; Aquilion, Toshiba, Tokyo, Japan). Conventional contrast-enhanced CT examinations were performed using 2 mL/kg of nonionic contrast material at a rate of 1.5 mL/s with a 120-s scanning delay. CT angiography examinations were performed using 100 mL of nonionic contrast material at a rate of 4 mL/s, and scanning delay was determined using a bolus tracking method. Other parameters of CT angiography were 120 kVp, 125 mAs, and 0.5-s rotation time. CT urography was performed using a 300-s scanning delay.

Two radiologists each with more than 12 years'

experience interpreting CT images, reviewed axial MDCT images with a 1- to 5-mm reconstruction interval on a picture archiving and communication system workstation. The narrowest part of the connection between the halves of the kidney was considered the renal isthmus (Fig. 1).

For divide fusion site the isthmus, those radiologists measured a maximum diameter of the outline of the HSK on the axial image and decided the center of the kidneys. Midline fusion was classified the position of the minimum width of the renal isthmus was less than 5mm on both sides from the center (Fig. 2). They independently measured minimum width of

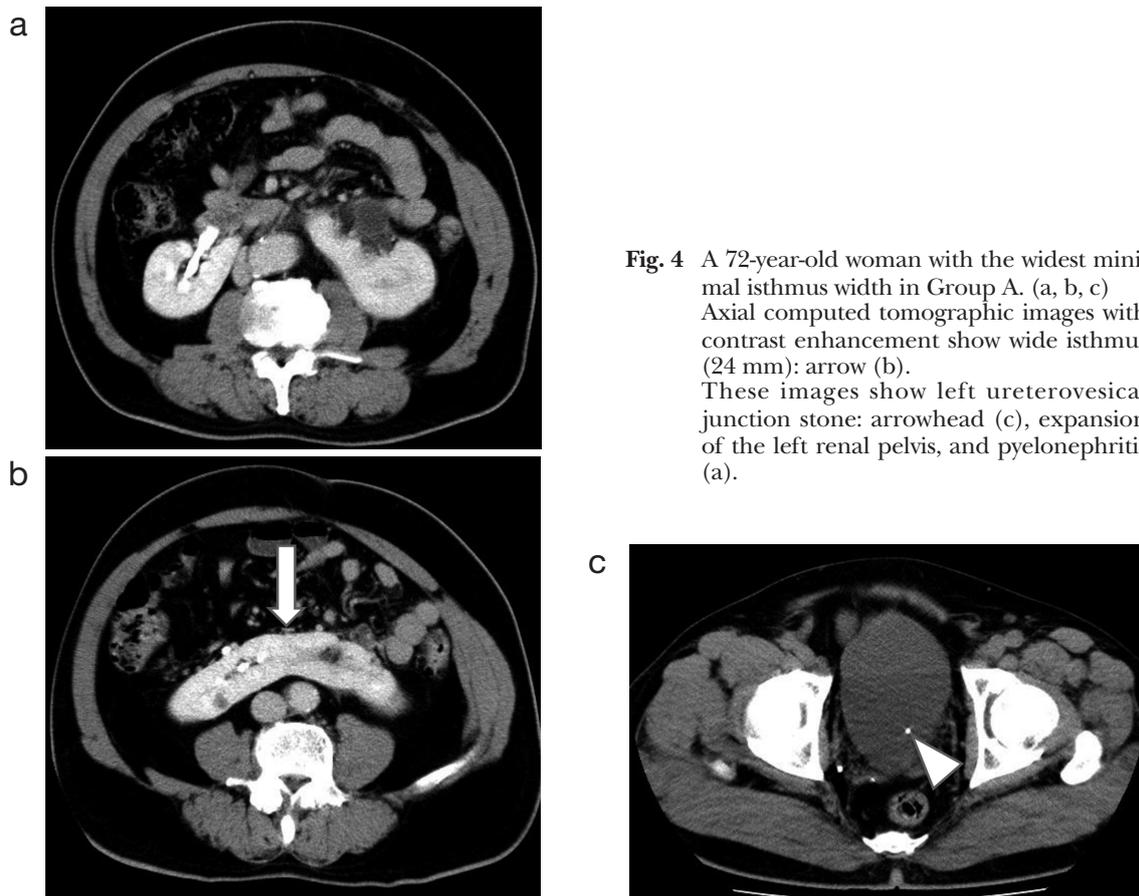


Fig. 4 A 72-year-old woman with the widest minimal isthmus width in Group A. (a, b, c) Axial computed tomographic images with contrast enhancement show wide isthmus (24 mm): arrow (b). These images show left ureterovesical junction stone: arrowhead (c), expansion of the left renal pelvis, and pyelonephritis (a).

Table 1 Renal function and renal shape.

	group A	group B	p
Minimum width of isthmus (mm)	11.0 ± 5.6	9.5 ± 5.1	P = 0.048
Renal function			
Cr. (mg/dl) [0.6-1.1]	1.22 ± 1.8	1.04 ± 1.0	P = 0.502
eGFR (ml/min./1.73m ²) [≥ 90]	70.1 ± 38.7	68.0 ± 27.2	P = 1.14
BUN (mg/dl) [9-21]	19.9 ± 11.6	17.6 ± 12.8	P = 0.067
Maximum length of pelvis (mm)			
Right side	9.3 ± 6.0	9.8 ± 8.9	P = 0.531
Left side	11.7 ± 10.0	9.3 ± 5.8	P = 0.517
Max size	14.3 ± 9.7	11.9 ± 8.8	P = 0.270

Cr.: creatinine; eGFR: estimated glomerular filtration rate; BUN: blood urea nitrogen; Max size: maximum length of pelvis on both sides

the renal isthmus and maximum length of the renal pelvis on each side on non-contrast enhanced axial CT image with 5-mm reconstruction in each patient (Fig. 3), and we used the mean value of each of those measurements by the 2 radiologists as measured value. We analyzed the correlation between isthmus width and renal pelvic length by linear regression analysis on both groups. We recorded renal function (creatinine: Cr., estimated glomerular filtration rate: eGFR and blood urea nitrogen: BUN). We compared demographic data, renal function, minimum isthmus width, and maximum renal pelvic length between groups using chi square test, student t-test, and Mann-Whitney U-test. Statistical significance was set at $P < 0.05$. Those 2 radiologists evaluated coexisting urological diseases excluding urolithiasis and neoplastic diseases diag-

nosed on non-contrast or contrast enhanced CT. The two resolved any disagreement through discussion to achieve consensus.

RESULTS

On all patients, two kidneys were connected by an isthmus at the lower poles. There was no rare variation of HSK with a common renal pelvis. There was 1 of lateral fusion which one of the pelvicalyceal systems drained a portion of renal tissue which extended across the midline [12]. Table 1 shows fusion site. Patients with midline fusion were 35 and patients with lateral fusion were 71, and left side fusion was more frequently than right fusion on both groups (Group A: 42, Group B: 23).

The overall mean minimum width of renal isthmus



Fig. 5 A 74-year-old woman with the largest renal pelvis, right ureteropelvic junction obstruction (UPJO), and pyonephrosis. (a) Axial and (b) coronal reformatted computed tomographic images with contrast enhancement show right severe hydronephrosis and pyonephrosis caused by UPJO and urinary tract infection.

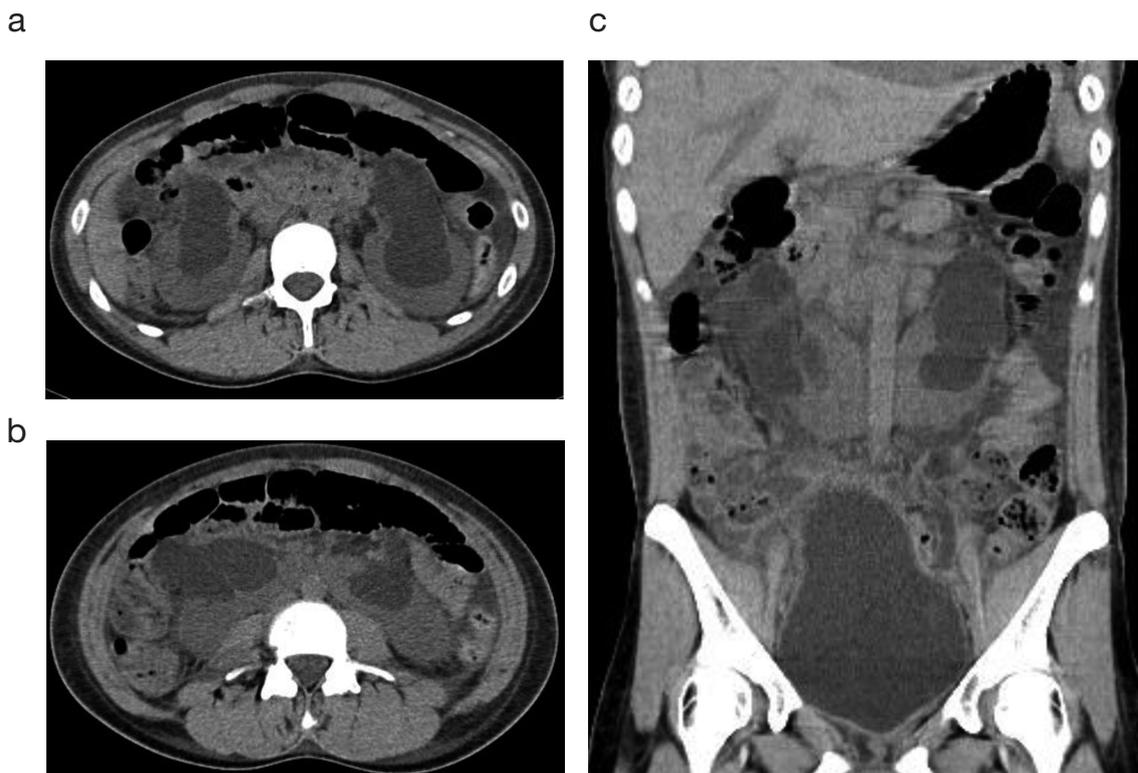


Fig. 6 A 20-year-old woman with neurofibromatosis type 1. (a, b) Axial and (c) coronal reformatted computed tomographic images without contrast enhancement show severe hydronephrosis, hydroureter, and neurogenic bladder. (a, c) The appearance of the renal pelvis opening is consistent with horseshoe kidney.

was 10.0 ± 5.3 mm; in 3 patients, the isthmus consists of thin fibrous band which was less than 1 mm. Minimum width of isthmus was larger in Group A than B ($P = 0.048$). And the largest minimum width of isthmus, 24 mm, was in Group A (Fig. 4).

Mean minimum width of renal isthmus was $8.4 \pm$

5.1 mm on midline fusion patients and it was 10.7 ± 5.9 mm on lateral fusion patients. There was no significant difference of minimum width of renal isthmus on fusion site ($P = 0.078$).

The overall mean maximum length of the renal pelvis was 12.7 ± 9.2 mm, and maximum renal pelvis

Table 2 Frequencies of coexisting diseases.

	group A	group B
Urological disease without urolithiasis	61.1% (22/36)	45.7% (32/70)
Neoplasm*	13.9% (5/36)	21.4% (15/70)
Others	8.3% (3/36)	11.4% (8/70)

*Non urological neoplasm in both groups

Neoplasm: pancreatic ca. colorectal ca. gastric ca. prostate ca. HCC, malignant lymphoma etc.

Table 3 Coexisting urological and neoplastic diseases.

	group A	group B
Urological disease	36 patients	32 patients
Renal cyst	15	20
Hydronephrosis	21	39
Pyelonephritis	6	2
Renal atrophy	1	1
UPJO	2	1
Renal injury	0	1
Renal infarction	1	3
Neurogenic bladder	0	1
Bladder carcinoma	0	1
Total	82	69

Neoplasm	5 patient	15 patients
Pancreatic cancer	1	1
Colorectal cancer	0	6
Gastric cancer	0	3
Gall bladder cancer	1	0
HCC	1	1
Cervical cancer	1	0
Prostate cancer	1	0
IPMN	0	2
Malignant Lymphoma	0	2
Liver metastases	0	1
Malignant peritonitis	0	1
Total	5	17

HCC: hepatocellular carcinoma; IPMN: intraductal pancreatic mucinous neoplasm; UPJO: ureteropelvic junction obstruction. Total number of diseases exceeded the number of patients because some patients had more than 2 diseases.

length did not differ significantly between Groups A and B ($P = 0.27$). UPJ obstruction caused the largest maximum renal pelvis length (65 mm) (Fig. 5). One patient in Group A and one in Group B had hydronephrosis with renal atrophy. In Group A, Hydronephrosis (dilatation of 10mm and more of renal pelvis) in 7 of the 12 patients was caused by ureteral stones. In one of those patients with hydronephrosis and renal atrophy in Group B, the youngest patient in both groups, neurogenic bladder was associated with neurofibromatosis type 1 (Fig. 6); another patient had left-side hydronephrosis as a result of renal hilar lymph node metastasis from pancreatic cancer.

There was no relationship between minimum isthmus width and maximum renal pelvis on both groups ($R^2=0.4$).

Demographic data did not differ significantly between Groups A and B (age, $P = 0.46$; sex, $P = 0.46$).

Table 2 shows renal function (Cr., eGFR and BUN) and renal shape (minimum width of isthmus and maximum length of renal pelvis) findings for both groups. Overall mean Cr. was 1.10 ± 1.32 mg/dL, mean eGFR was 68.8 ± 31.4 mL/min/1.73 m² and mean BUN was 18.4 ± 12.4 mg/dL. Neither Cr., eGFR nor BUN did not differ significantly between Groups A and B ($P = 0.502$, $P = 1.148$, $P = 0.067$). The level of Cr. was normal (< 1.0 mg/dL) in 80 patients (80/106, 75.5%), eGFR was normal (≥ 90) in 15 patients (15/106, 14.2%) and BUN was normal (9 to 21 mg/dL) in 82 patients (82/106, 77.4%).

Table 3 shows frequencies of coexisting diseases and coexisting urological diseases excluding urolithiasis

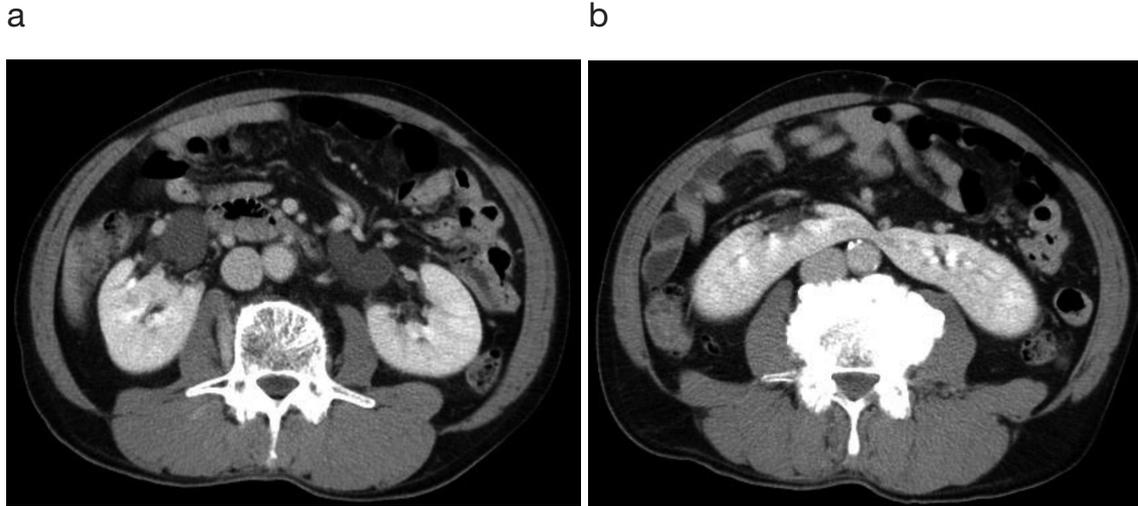


Fig. 7 A 65-year-old man with expansion of both sides of renal pelvis. (a, b) Axial computed tomographic images with contrast enhancement show bilateral dilation of the renal pelvis (right, 23 mm; left, 24 mm) of unknown cause.

and neoplastic diseases. There was no renal malignant tumor in either groups.

DISCUSSION

HSK is a well-known congenital anomaly of the upper urinary tract [1, 2]. HSK results from the fusion of metanephric buds between weeks 4 and 8 of embryogenesis, which blocks their cephalic migration and normal rotation [1, 3, 7, 9]. The inferior mesenteric artery prevents the cranial migration of the isthmus of the fused kidney, which remains in the lower part of the abdomen [1, 7, 9]. Typically, the urinary collection system is anteriorly displaced, with lying anterior to the isthmus [1-7]. When fusion is in the midline, the position of the renal pelvis depends on the width of the isthmus [7], which comprises normal kidney tissue or fibrous band. The developing kidneys are connected by an isthmus containing true renal parenchyma when abnormal fusion results from the aberrant migration of posterior nephrogenic cells [13]. Alternatively, HSK results when median fusion of the metanephric tissues during an early embryologic stage produces a fibrous isthmus [14]. In 15% of HSK, the isthmus is fibrous; [15] the isthmus was fibrous in three of our patients (2.8%). Moreover, the renal pelvis is often large, flabby, and extrarenal, and the ureter inserts abnormally high in the pelvis [1, 7]. In our study, mean maximum length of the renal pelvis was 12.7 ± 9.2 mm, and maximum renal pelvis length did not differ significantly between patients with HSK with or without urolithiasis. Mean renal pelvis length of 12.2 to 13.3 mm has been reported and is consistent with our results [3]. UPJ obstruction accounted for the longest maximum renal pelvis length (65 mm) (Fig. 5), but the cause of ureteral stenosis was unclear in a patient with maximum length of renal pelvis about 24 mm (Fig. 7). Therefore, we considered that ureteral stenosis does not necessarily accompany dilation of the renal pelvis in patients with HSK.

Glodny's group [3] offers the only report about the shape of HSK on CT evaluation. Because fusion occurs at the midline in about half of HSKs, they

measured the width of each side from the fusion site and reported width at the right of 16.3 mm and at the left of 34.4 mm on axial CT image [3]. They also evaluated isthmus location, length of the fusion site longitudinally, and renal rotation and argued against the rule that rotation of the renal axis depends on whether craniocaudal length of isthmus is more or less than a third of renal length [3, 12]. However, they did not assess differences of clinical significance associated with renal shape.

More than half cases had left side fusion in our study. Because the isthmus is in front of the abdominal aorta and abdominal aorta laid on the left side of the vertebra in many cases, left side fusion may be occurred frequently.

Many HSKs are asymptomatic and found incidentally. In our study, most patients demonstrated normal renal function, and about 69.4% of patients with HSK with urolithiasis had no symptoms. However, a wide variety of associated congenital and acquired genitourinary and nongenitourinary anomalies are associated with HSK [2-10, 15-17]. High insertion of the ureter into the renal pelvis in up to 35% of cases causes varying degrees of stenosis of the UPJ that may often cause problems [5, 6]. Stone diseases are also common in HSK. Kidney stones develop in 20 to 60% of patients and are associated with obstruction and recurrent infections [5]. Urinary stasis and urolithiasis also predispose HSK to infection, which occurs in 27 to 40% of patients, in our study, 36 of 106 adult patients (34.0%) had urolithiasis, incidence similar to that previously reported [18]. To our knowledge, however, the relationship between renal shape and stone formation has not been evaluated on ultrasonography (US) or CT examination. The isthmus may not be visualized on US for several reasons [11]. In approximately 15% of fused kidneys, the fibrous connecting band may not be visible on US. As well, the isthmus is usually situated low in the abdomen, at the level of the L4-5 vertebral bodies, an area frequently obscured by bowel gas [11]. The isthmus may also be mistaken for normal anatomic structures, such as the pancreas, or for pathologic

retroperitoneal conditions. We speculated that malrotation with anterior displacement of the collecting system, narrowing of the renal hilum, and superior and lateral ureteric insertion into the renal pelvis were severe in cases of HSK with wide isthmus and that wide isthmus was a risk factor of stone formation. Because CT is useful for the accurate determination of stone location and volume and is used routinely prior to treatment in patients with HSK, we evaluated width of the isthmus on CT. In our study, minimum isthmus width of patients with HSK was larger in those with urolithiasis (11.0 ± 5.6 mm) than without urolithiasis (9.5 ± 5.1 mm). As expected, these results showed that wide isthmus was a risk factor for stone formation.

Treatments of patients with HSK with urolithiasis include percutaneous or laparoscopic nephrolithotomy, extracorporeal shock wave lithotripsy, and surgery [5, 6]. Surgical treatments of patients with HSK with urolithiasis and UPJ obstruction are pyeloplasty and laparoscopic or open hemi/total nephrectomy [18]. The choice of treatment depends on stone size/location and the appearance of the pelvicalyceal system on preoperative imaging [5, 6, 18]. Particularly, presurgical evaluation of the collecting system and isthmus is important, and contrast-enhanced CT including CT angiography is essential for preoperative evaluation of abnormal anatomy because of the complexity of the isthmus blood supply [19, 20].

Patients with HSK have an increased incidence of renal tumor, such as Wilms, renal pelvic, and carcinoid tumors [3, 7, 16, 17]. Renal cell carcinoma is the most common neoplasm in these patients but does not occur more frequently than in the general population [16, 21]. The incidence of neoplasm in HSK is estimated to be one to 12%, although this likely represents the upper limit, in view of the potential reporting and selection bias [16, 21]. None of our study patients had associated renal tumor, but the population of our study was small. The incidence of tumors of the renal pelvis is approximately 3 to 4 greater in patients with HSK than with normal kidney configuration, and squamous cell carcinoma is more common than urothelial carcinoma [16, 17, 21]. The high incidence of squamous cell carcinoma is related to chronic obstruction, urolithiasis, and infection, common complications of HSK [16, 17]. In this study, wide isthmus is related to stone formation. Because urolithiasis is one of the risk factor of renal pelvic carcinoma, special attention should be paid to width of isthmus on CT.

Our study has several limitations. Our sample was small and included only adults. We evaluated width of isthmus only on axial CT images. And statistical difference of incidence of urolithiasis between patients with wide and thin isthmus was small ($P = 0.048$). Error of measurement on only axial CT image might have an influence on the results. We did not perform metabolic analysis of HSK with urolithiasis such as serum calcium level nor urinary calcium excretion. Multiplanar images and 3-dimensional assessment are needed to assess exact renal shape including renal rotation. A role of metabolic abnormalities is reported in stone formation in patients with HSK [24], and we suspect anatomic anomaly as a risk factor for metabolic irregularity. A large-scale study of patients of all ages is

needed to investigate correlations between stone formation, renal shape, and metabolic analysis in patients with HSK. Nevertheless, CT is used routinely in these patients, and evaluation of renal shape on axial image is convenient.

In conclusion, we evaluated the relationship between urolithiasis and characteristics of renal shape of HSK on MDCT in adult patients and patients of HSK with wide isthmus might have a tendency of a high incidence of stone formation. Because urolithiasis, urinary stasis, and infection are risk factors for tumor of the renal pelvis, it is useful to measure the width of isthmus on axial CT images and attention is more necessary for the progress observation of HSK patients with wide isthmus.

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