

Double inferior vena cava with three shunts: a rare anomaly with important implications for surgeons

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Inferior vena cava (IVC) is the largest single vein that collects systemic venous blood from the lower part of the body except the gut and drains into the right atrium. Double IVC is a rare anomaly in humans and usually is discovered incidentally during the interventional radiological procedures or routine cadaveric dissection. Here we report a rare case of unusual observations in an adult female Thai cadaver with a duplicated left IVC with three short venous shunts and a variant pattern of the hemiazygos vein. Also included in this case was the presence of unilateral double renal vein on the right kidney. This type of anatomic variation of the great vein has never been reported before. A detailed description of these variations is useful and essential for the surgeons during approaching the retroperitoneal region. (Folia Morphol 2017; 76, 2: 307–311)

Key words: inferior vena cava, supracardinal veins, venous shunt

INTRODUCTION

In human, the inferior vena cava (IVC) is typically a single vein located on the posterior abdominal wall in the retroperitoneal space of the abdomen. It is formed by the union of the right and left common iliac veins in front of the 5th lumbar vertebra. This largest vein and its tributaries normally return deoxygenated blood from the lower part of the body except the gut. It then ascends cranially along the right side of the vertebral column with the aorta running laterally on the left and passing through the diaphragm to drain into the right atrium on its posterior side [23]. Variations of IVC may occur due to many complicated transformation processes during embryonic life [15, 19]. Anatomical variation of the

IVC occurs in 0.4–4% of the population [14]. Among the abnormalities related to the formation of the IVC, the double IVC (DIVC) was the most common anatomic variation [3]. We report here a case of DIVC with three short venous shunts and a variant pattern of the hemiazygos vein in a Thai cadaver. The relevant literature, the embryogenesis of the IVC related to this rare anatomical variant, and the clinical significance were reviewed and discussed.

CASE REPORT

During routine anatomical dissection for medical students in the Department of Anatomy, Faculty of Medicine, Khon Kaen University, an anatomical variation of the IVC with associated variations were

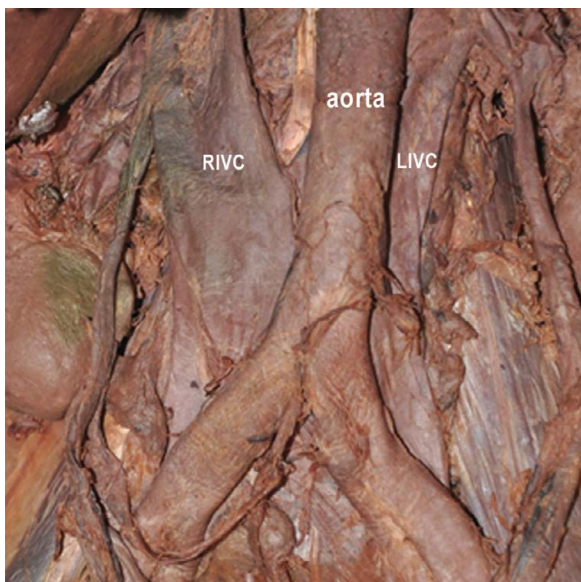


Figure 1. Dissection of retroperitoneal region showing the anatomical variation of double inferior vena cava on both sides of the abdominal aorta; RIVC — right inferior vena cava; LIVC — left inferior vena cava.

observed in a 45-year-old Thai female cadaver. The retroperitoneal region including all of the vessels concerned was carefully dissected. The measuring instrument used was sliding Vernier calliper.

This dissection was approved by the Human Ethics Committee of Khon Kaen University, Khon Kaen, Thailand (Approval No. HE 571314).

OBSERVATIONS

In the retroperitoneal region, DIVC was detected unexpectedly on both sides of the abdominal aorta (Fig. 1). The abdominal aorta was cut and reflected to expose the anomalous venous channels (Fig. 2A). All the anomalies were presented by outline drawing in Figure 2B for the clarity.

At the confluence of the left internal iliac and external iliac veins, a duplicated left IVC was found. Its dimensions were 12.73 cm in length and 0.68 cm in diameter. It passed upward posterior to the left side of the abdominal aorta to join the left renal vein (Fig. 2). After receiving blood from the left renal vein, the left IVC continued to become the hemiazygos vein. It travelled upward medially passing in front of the body of the 8th thoracic vertebra to join the azygos vein at the right side of the intervertebral disc between the 7th–8th thoracic vertebrae. The azygos vein then received the superior intercostal veins before terminating into the right atrium. The left suprarenal vein

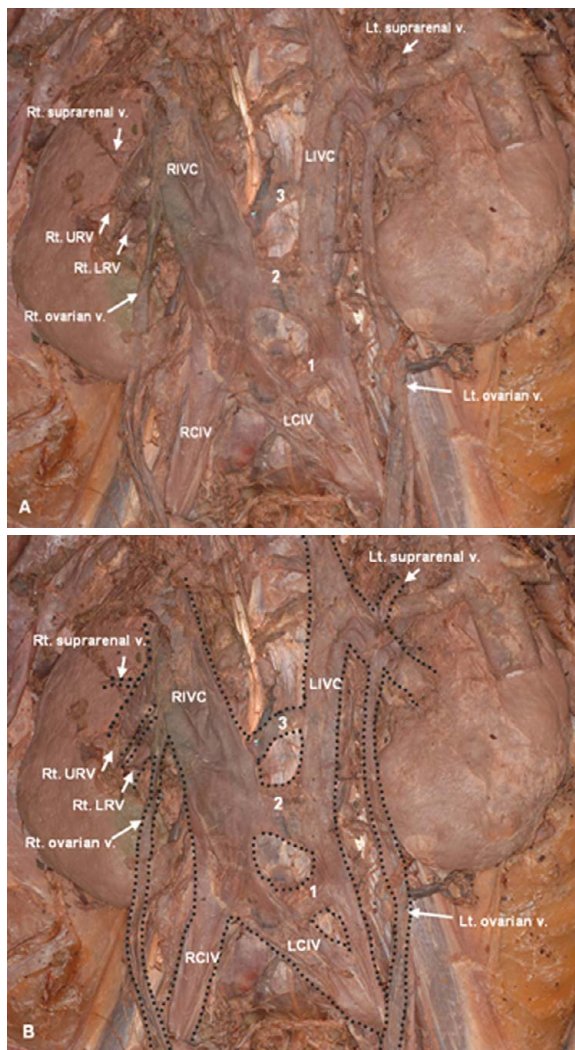


Figure 2. A. Double inferior vena cava and the three short venous shunts. The double right renal veins emerged from the hilum of the kidney to enter the right inferior vena cava. The right suprarenal renal vein drained into the right upper renal vein. The right ovarian vein drained into the junction of the right lower renal vein and then to the right inferior vena cava; **B.** Outline drawing of the anatomical variations of the panel A; URV — upper renal vein; LRV — lower renal vein; RCIV — right common iliac vein; LCIV — left common iliac vein; RIVC — right inferior vena cava; LIVC — left inferior vena cava.

and the left ovarian vein drained into the left renal vein as usual.

The right IVC was formed by the joining of the left and right common iliac veins as usual but its route deviated from the centrally lined abdominal aorta when running up to the right kidney. The length of the right abdominal IVC was 13.5 cm and the diameter was 1.4 cm.

Three short venous shunts were noticed (Fig. 2A, B). The 1st shunt was 1.92 cm in length and connected the left common iliac vein and the left IVC. The 2nd

and 3rd shunts, 1.67 cm and 1.25 cm in length, respectively, connected the right and the left IVC.

Other associated anomalies were the two right renal (upper and lower) veins merging from the renal hilum to enter the right IVC. The right suprarenal renal vein drained into the right upper renal vein whereas the right ovarian vein drained into the venous angle formed by the right lower renal vein and the right IVC.

DISCUSSION

The DIVC was first described in a male cadaver by Lucas [13]. The prevalence of DIVC is estimated between 0.2% and 3% [4].

Ang et al. [1] reviewed and analysed 41 published articles about DIVC between 2000 and 2011 and found that there were 53 (31 males, 21 females, and 1 unknown) cases of DIVC from 18 countries and the incidence of DIVC was slightly higher in males than in the females. In addition, we have found 5 more DIVC cases in the same period of 2000–2011, which have not been included in the review of Ang et al. [1]. Artico et al. [3] detected a case of DIVC in an adult male patient during a computed tomography (CT)-scanning for follow-up of pancreatic cancer. Kumar [10] reported a case of an adult male cadaver having the DIVC, which ascended to join the left renal vein before connecting to the right IVC. Likewise, Xue et al. [24] found a Japanese male cadaver, having a DIVC, in that the left IVC coalesced with the left renal vein and finally ran across in front of the abdominal aorta and drained into the right IVC. Two cases of DIVC were found during renal transplantation: Kennealey et al. [9] found a DIVC in a female living renal transplant donor and Raza et al. [17] found a DIVC in a male deceased organ donor. Discovery of the anomalous vessels during transplantation caused prolongation of the operative time to re-design the procedure for safety outcomes.

After extensive literature survey, we have found 13 additional cases (6 males and 7 females) of DIVC from 2012 to 2015: A case of DIVC was found during phlebography for the treatment of a pulmonary thromboembolism from multiple traumas in a male patient [18]. Loo and Jhummon-Mahadnac [12] reported 2 cases of duplicated left IVC in Australian, 1 case was a female cadaver and the other case a living adult male who underwent a staging CT-scan for preoperative work-up. In both cases, duplicated left IVC began at the left common iliac vein, joining the left renal vein, and then crossed the abdominal

aorta anteriorly to enter the right IVC. Hayashi et al. [7] reported 3 cases of DIVC (2 males and a female) in Japanese cadavers and described three types of the flow pattern of DIVC. In addition to the literature review of 2000–2011, Ang et al. [1] reported 3 more cases of DIVC in female patients investigated by abdominal CT scan in New Zealand. Tanka [22] reported 2 cases (1 case each of male and female) of DIVC in Albania while CT imaging of the abdominal region. More recently, Kumar et al. [11] reported an incidental finding of DIVC in an Indian female patient on CT abdominal angiography. Furthermore, Polguy et al. [16] detected a case of DIVC by CT examination in a male patient with endovascular repair for abdominal aortic aneurysm. This finding was similar to the present case in that the left renal vein joined with the left IVC and continued as the hemiazygos vein. As can be seen in those literature surveys, more variations of the IVC have been increasingly detected along with the advance of many vascular imaging techniques.

Our DIVC case reported here is unique and different from all of the cases mentioned above, in that the left side IVC continued to become the hemiazygos vein after joining the left renal vein and the 3 shunts. Double right renal vein and the 3 shunts associated with DIVC have never been reported before.

Although the clear aetiology of DIVC is still unknown, understanding of the complicated embryogenesis of the venous system is helpful to understand the vascular variation reported here. The IVC is formed by regression, anastomosis and replacement of the foetal venous blood system during 4th–8th week of gestation [8, 19, 21]. Initially, there were composed of the three sets of paired veins; the posterior cardinal veins, the subcardinal veins, and the supra cardinal veins, which are formed at the retroperitoneal region. The posterior cardinal veins primarily collect the caudal half of venous blood in the earliest embryo development and then regress. During the 5th and 8th week, while the posterior cardinal veins gradually relapse, the bilateral subcardinal veins develop and become dominant at 7th week. By the 8th week, the posterior cardinal veins become gradually degenerated, and then superseded by the supracardinal veins [8]. Numerous anastomotic vessels connect the each side of subcardinal veins and posterior cardinal veins including the supracardinal veins. As the subcardinal veins remodelled, the posterior cardinal veins obliterated. The posterior cardinal veins degenerate almost completely and remain only

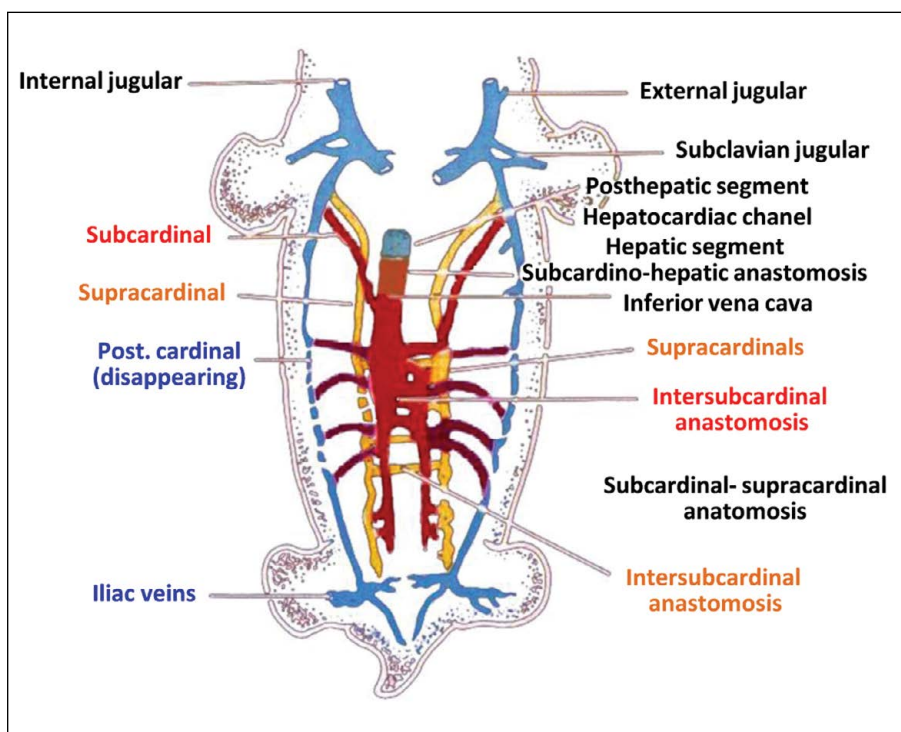


Figure 3. Remodelling and anastomosis of the three sets of primordial paired veins during embryogenesis to yield the final composition of the inferior vena cava (reproduced from Figure 4, In: Kellman et al. 1988; with permission from The Radiological Society of North America, RSNA).

as the common iliac vein and the caudal most sacral portion of the IVC. The right supracardinal vein anastomoses with the right subcardinal vein to form an infrarenal segment of the IVC whereas the lower portion of the left supracardinal vein obliterated as showed in Figure 3 [8]. Therefore, the double IVC observed in the present case might have resulted from the persistence of the caudal part of both left and right supracardinal veins. The 1st venous shunt connected between the left common iliac vein and the left IVC in the present case might be a remnant of the venous anastomosis between the left supracardinal vein and the left posterior cardinal vein. In contrast, the persistence of the intersupracardinal anastomosis during remodelling may yield the 2nd and 3rd shunts in the present case.

The occurrence of DIVC is generally asymptomatic, the presence of this aberrant venous anatomy may have significant clinical implications to the radiologists, surgeons, etc. The potential for misdiagnosis on imaging may occur, leading to unnecessary treatment [2]. The surgeons should be aware of such variation of the IVC when performing surgical procedures in retroperitoneal region such as laparoscopic nephrectomy or donor nephrectomy [6, 20]. When

attempting cava filtration, DIVC patient may require placement of a caval filter into each vessel in order to avoid the embolism [5]. DIVC is also a risk factor for formation of thrombosis back pain and anomalous circulation of blood to the heart as a consequence of changes in blood flow [21]. In addition, the DIVC can increase a risk of vascular injury during retroperitoneal procedures leading to life-threatening [6, 9]. Therefore, this asymptomatic anomaly has to be known prior to precede any abdominal surgery and sufficient preoperative planning is required to avoid an unexpected iatrogenic complication during the operation.

CONCLUSIONS

In conclusion, this is the first report of anatomic variation of DIVC with 3 shunts, associated with the continuation of left IVC to become the hemiazygos vein and unilateral double renal veins. These congenital variants caused by an unusual embryological development, are essential for clinical manipulation particularly for the surgeons during approaching the retroperitoneal region and the radiologist to avoid misdiagnosis as pathologic appearance on CT imaging.

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