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Case Report

Duplicated Inferior Vena Cava in a 69-Year-Old White Female Donor

Joanna Klansek [1], Keiko Meshida [2], Elizabeth Maynes [3], Maria Ximena Leighton [3], Gary Wind [3] and Guinevere Granite [3,*]

[1] F. Edward Hebert School of Medicine, Uniformed Services University of Health Sciences, Bethesda, MD 20814, USA
[2] The Henry M. Jackson Foundation for the Advancement of Military Medicine, Inc., Bethesda, MD 20814, USA
[3] Department of Surgery, Uniformed Services University of the Health Sciences, Bethesda, MD 20814, USA
* Correspondence: guinevere.granite@usuhs.edu; Tel.: +1-301-295-1500

Abstract: While relatively uncommon, a duplication of the inferior vena cava is moderately well-discussed in the literature. This anatomical variation was noted in a 69-year-old white female donor. This variation is typically asymptomatic; however, it can be associated with complications, such as confusion with a mediastinal mass, increased risk for thromboembolism, and hemorrhage during surgery. It is also associated with a handful of comorbidities, including, but not limited to, congenital renal anomalies such as horseshoe kidney or fused crossed kidney. Research supports that the variation of a duplicated IVC (DIVC) can be due to a failure of the left supracardinal vein to regress during embryonic development.

Keywords: duplicated inferior vena cava; venous embryonic development; supracardinal veins; anatomical variation; inferior vena cava variations

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1. Introduction

Malformations and/or variations in venous vasculature may affect any number of structures. They can be classified as extratruncular or truncular based on when they occur during embryonic development [1]. Lee (2012) describes the distinction as extratruncular variations occurring during early embryogenesis and truncular variations occurring later in life [1]. In general, truncular lesions are limited to a vessel trunk, and as such, may have more serious consequences for patients. Lee (2012) also states that truncular venous malformations can be the result of a persistent fetal remnant vein that did not involute or regress normally, which correlates with the variation presented in this case report (Figures 1a,b and 2) [1]. Throughout embryonic development of the inferior vena cava (IVC), there is a cycle of development and regression of venous pairs. These three venous pairs include the postcardinal, supracardinal, and subcardinal veins [1]. The segments must fuse and develop in proper chronology, which affords many opportunities for anomalies [1]. Failure of the left supracardinal vein to regress leads to a duplication of the IVC. Venous embryological anomalies may be associated with additional anomalies, such as renal anomalies, including horseshoe kidney and crossed fused kidney. Even though many of these anomalies, including a DIVC, typically present asymptomatically for the majority, if not all, of a patient's life, increased for thromboembolism and side effects of incidentally found comorbidities are essential to identify prior to any sort of abdominal surgery in order to improve surgical outcomes.

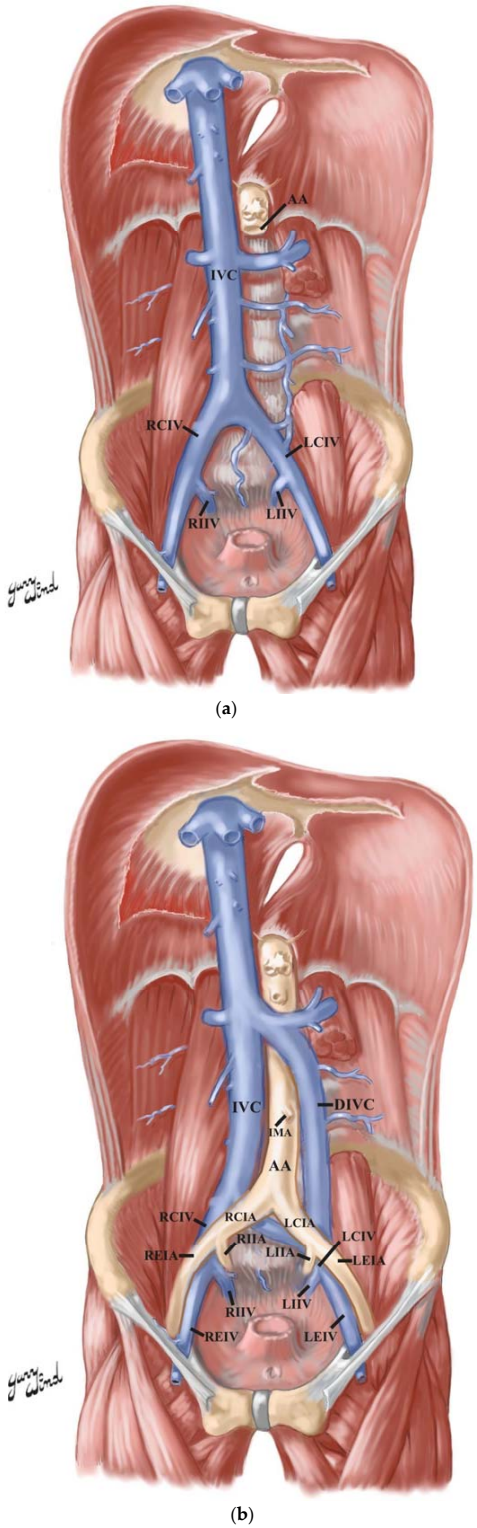

Figure 1. (**a**) Illustrative schematic of normal anatomy and tributaries of the inferior vena cava. (**b**) Illustrative schematic of a duplicated inferior vena cava and its tributaries. Here you can see the duplicated IVC draining into the left renal vein, then merging with the IVC. AA = abdominal aorta; DIVC = duplicated inferior vena cava; IMA = inferior mesenteric artery; IVC = inferior vena cava; LCIA = left common iliac artery; LCIV = left common iliac vein; LEIA = left external iliac artery; LEIV = left external iliac vein; LIIA = left internal iliac artery; LIIV = left internal iliac vein; RCIA = right common iliac artery; RCIV = right common iliac vein; REIA = right external iliac artery; REIV = right external iliac vein; RIIA = right internal iliac artery; RIIV = right internal iliac vein.

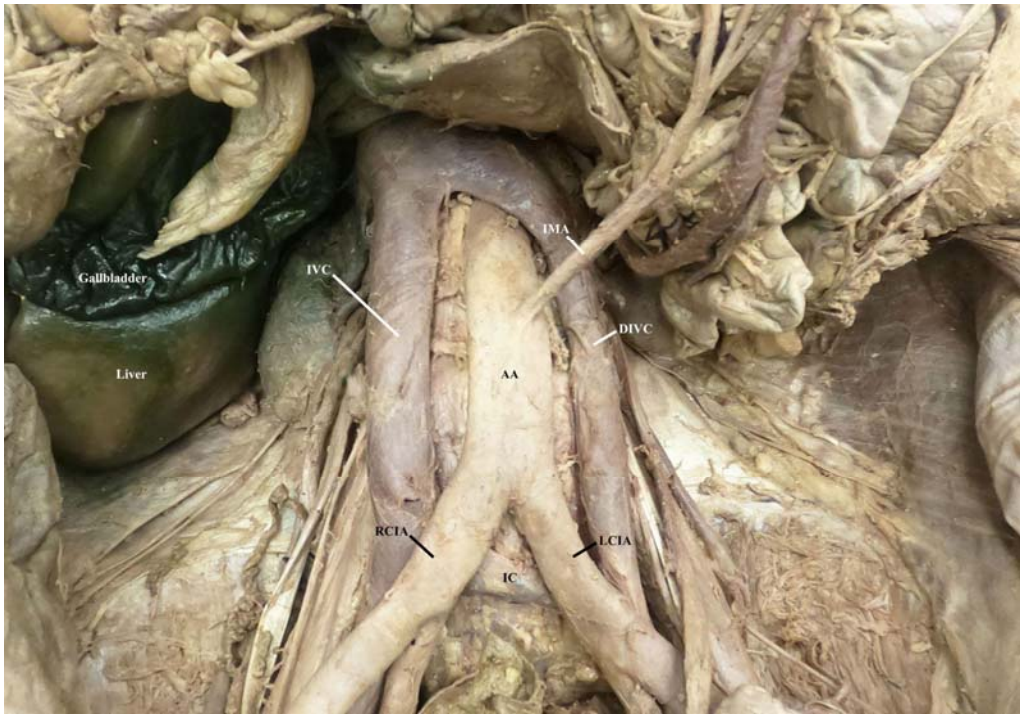

Figure 2. Facilitated display highlighting the duplicated inferior vena cava merging with the inferior vena cava anterior to the abdominal aorta and superior to the inferior mesenteric artery. Also highlighted is the additional venous connection at the base of the duplicated inferior vena cava and the inferior vena cava near the iliac veins. AA = abdominal aorta; DIVC = duplicated inferior vena cava; IMA = inferior mesenteric artery; IC = inferior connection (between the main and duplicated IVC); IVC = inferior vena cava; LCIA = left common iliac artery; RCIA = right common iliac artery.

2. Case Description

During routine anatomical dissection of sixty-five human cadaveric donors during the 2021 first-year medical gross anatomy course and 2021 graduate nursing advanced anatomy course at the Uniformed Services University of the Health Sciences, a duplicated IVC was observed in a 69-year-old white female donor with a listed cause of death of Corticobasilar Degeneration (Neurodegenerative Disease). There was an additional venous connection at the base of the two IVCs near the iliac veins (Figure 2). All cadaveric images depicted in this article were vetted and approved by the USUHS Human Anatomical Specimens Review Committee (HAMRC).

3. Discussion

3.1. Duplicated or Double Inferior Vena Cava

In the majority of cases, the IVC directs deoxygenated blood flow upwards from the inferior and middle body, draining into the right atrium. To achieve this goal, the IVC is located on the right side of the body (Figure 1a). In comparison, a person with a duplicated/double IVC has an IVC on both the left and right sides of the body (Figure 1b). To drain deoxygenated blood into the right atrium of the heart, the left IVC must cross over the abdominal aorta (AA) and merge with the right IVC. This is demonstrated in this particular case as the two IVCs merge anterior to the AA and superior to the inferior mesenteric artery (IMA) (Figure 2). Prior to merging directly inferior to the right atrium, the left and right IVCs drain from the left and right iliac veins (Figure 3). There is a second merge between the left and right common iliac veins posterior to the AA and inferior to the aortic bifurcation into the left and right common iliac arteries, creating a rectangular venous structure in the abdomen (Figures 2 and 3). Measurements of the DIVC were conducted through ImageJ software, using a baseline of 4.10 mm for the diameter of the IMA at its

origin, according to a study conducted by Sinkeet et al. (2012) [2]. Four measurements of the DIVC width were conducted along the vessel to average an estimated width of 11.83 mm. To measure the length, the outer and inner lengths were computed to be 11.71 cm and 10.12 cm, respectively, averaging an estimated DIVC length of 10.91 cm.

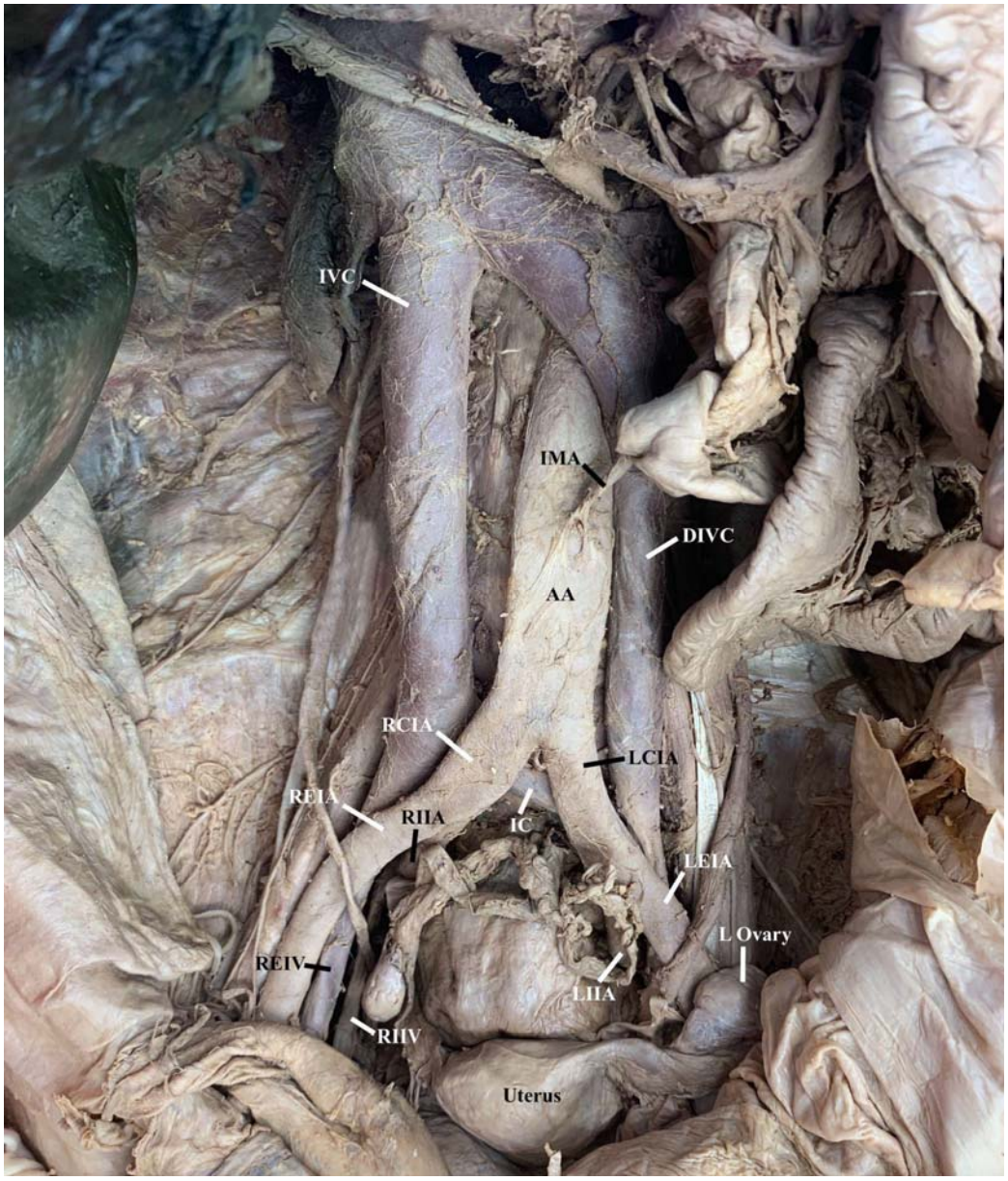

Figure 3. Facilitated display highlighting the second merge of the duplicated inferior vena cava and the inferior vena cava posterior to the abdominal aorta and inferior to the aortic bifurcation between the left and right common iliac veins, creating a rectangular venous structure in the abdomen. AA = abdominal aorta; DIVC = duplicated inferior vena cava; IMA = inferior mesenteric artery; IC = inferior connection (between the main and duplicated IVC); IVC = inferior vena cava; LCIA = left common iliac artery; LEIA = left external iliac artery; LIIA = left internal iliac artery; RCIA = right common iliac artery; REIA = right external iliac artery; REIV = right external iliac vein; RIIA = right internal iliac artery; RIIV = right internal iliac vein.

3.2. Embryonic Development

The IVC develops between weeks 6 and 8 of embryonic development [3]. During these three weeks, three pairs of veins emerge: postcardinal veins, subcardinal veins, and

supracardinal veins (Figure 4) [1]. Regression of the left supracardinal vein allows for the right-sided nature of the mature IVC in the majority of humans. Failure of this regression can result in a duplicated IVC, as noted in this donor [1]. If the right supracardinal vein regresses instead of the left, this can present as a circumcaval ureter, where the ureter passes posterior to the IVC [4]. Circumcaval ureters are clinically significant because they can lead to renal complications such as ureteral obstructions and hydronephrosis [4].

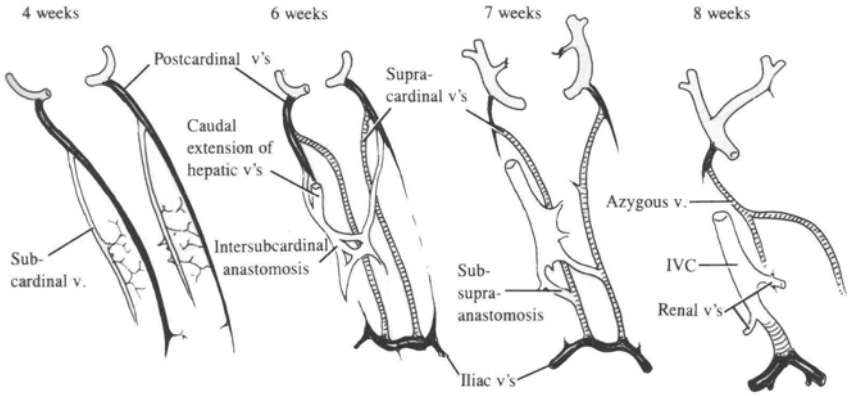

Figure 4. Embryological schematics created by Dr. Gary Wind illustrating the abdominal venous system at four weeks, six weeks, seven weeks, and eight weeks.

3.3. Associated Comorbidities

Investigating other literature reviews of this rare anomaly revealed multiple possible interesting comorbidities. For example, Shaha et al. (2016) documented a case report of a patient who presented with DIVC, crossed fused kidney, and gut malrotation [5]. A possible explanation for these comorbidities would be adjustments to the body's vasculature during embryological development. In particular, renal relocation could be impaired by the presence of a second DIVC occupying abdominal space. This could be a possible explanation for the association with a crossed-fused kidney, where both kidneys are fused on one side of the abdomen, or a horseshoe kidney, where the kidneys are bound by their inferior poles. Both of these anomalies typically present asymptomatically; however, throughout the literature, they have been demonstrated to have associations with an increased risk of developing renal cancer. Renal biopsy for diagnosis of renal cancer is controversial and can indicate a nephrectomy based on imaging results alone. Even then, renal biopsy is usually contraindicated (unless extreme circumstances arise) for evaluation of chronic kidney disease (CKD) if a patient only has one kidney. This contraindication exists because if excessive bleeding were to occur during the biopsy, the ultimate treatment to stop the bleeding is a nephrectomy. In both horseshoe kidney and fused kidney, renal cancer may also involve a complete nephrectomy. In both of these situations, a nephrectomy would place the patient on permanent dialysis as they are no longer able to filter their own blood. Shaha et al. (2016)'s findings also cite sources that support associations between DIVC and polycystic kidney disease, horseshoe kidney, and ectopic kidney [5]. These associations with renal anomalies are important to discover prior to any abdominal surgery in order to preserve renal vasculature during surgery and particularly avoid the possibility of a nephrectomy of a horseshoe or fused kidney if at all feasible. Shaha et al. (2016)'s conclusions encourage surgeons to complete a thorough exploration for other congenital anomalies should a DIVC or other rare finding appear on routine examination or preoperative imaging [5].

Gut malrotation is also mentioned as a comorbidity in Shaha et al. (2016)'s article, although research indicates it is a less common comorbidity than renal malformations [5]. Intestinal malrotation may lead to volvulus, as the intestine fails to rotate correctly during embryological development. This situation requires urgent surgical correction. If not corrected, parts of the bowel may become ischemic and die. The association between gut

malrotation and fused crossed kidney both involve failure of proper positioning during embryogenesis. Their association with a duplicated IVC could possibly be explained by a mutation in gene signaling that leads to incorrect completion of embryogenesis, although further studies would be needed to investigate this hypothesis.

3.4. Clinical Significance

Duplicated IVCs are typically incidental findings and present asymptomatically. Research, however, supports a handful of associated complications and surgical relevance. For example, on imaging, the IVC duplication may appear as a mediastinal mass [6]. On computerized tomography (CT), this may be confused with an abdominal aneurysm or aortic dissection. This misinterpretation can lead a patient to undergo unnecessary tumor work-up and imaging [7]. Additionally, due to the increased surface area of vasculature, patients with a duplicated IVC have a higher risk of chronic congestion and hypertension [1]. Patients also present with increased risk for thromboembolism, as a thrombus could become lodged at the extra bifurcation. This could also lead to life-threatening pulmonary implications, such as pulmonary thromboembolism [3]. This risk for IVC thrombosis increases significantly for patients with any type of congenital IVC anomaly. Li et al. (2022) state a prevalence of 60–80% for these patients [8]. Taking into account Virchow's Triad of hypercoagulable state, stasis, and endothelial damage, increased venous stasis from insufficient blood return and drainage can explain this increased risk [8]. Treatment with an IVC filter can increase blood flow, but knowledge of the duplication is essential for proper placement [8]. In particular, Rao et al. (2011) suggest the placement of an IVC filter in both the left and right IVCs [9]. Filter placement may have its own complications, including filter tilting, fracture, perforation, or malposition [9]. These risks increase in filter placement in a DIVC as there are two filters to consider—one for each IVC. This addition makes imaging and precision that much more important. Surgically, knowledge of a DIVC is important prior to any abdominal procedure as it impacts access to neighboring organs and vasculature. Failure to acknowledge a DIVC in a patient could lead to unintentional hemorrhage and death [10]. For example, Effler et al. (1951) describe a potentially preventable fatality after ligation of the anomalous bilateral IVC [10]. This complication arose after an exploratory thoracotomy with right total pneumonectomy following identification of a bronchial neoplasm [10]. Failure to take note of the duplicated IVC after identifying the hepatic vein led to a total ligation of the anomalous IVC during surgery. Ligation resulted in massive retroperitoneal hemorrhage and, ultimately, the patient's death [10]. Identification of this vessel prior to the procedure and plans to reconstruct the vessel during surgery may have avoided the patient's outcome. CT is currently the gold standard for identifying duplication of the IVC and should be utilized prior to abdominal surgery.

4. Conclusions

During embryological development, persistence of both supracardinal veins can lead to a DIVC, as noted in this case. Although this particular patient died of corticobasilar degeneration, symptoms of mediastinal widening on CT and thromboembolism could have appeared if the patient was symptomatic. Associations with gastrointestinal or renal anomalies may have been present asymptomatically as well but were not noted in this particular case. Future research could explore possible genetic associations with such comorbidities and congenital anomalies; perhaps there is an association with a mutation in gene signaling. In conclusion, a DIVC is important to identify via CT in patients prior to undergoing abdominal surgery since it can pose a hemorrhagic threat as well as decreased visibility of left-sided vasculature and anatomy.

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