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Duplication of the inferior vena cava: evidence of a novel type IV

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Abstract: Anatomical variations of the inferior vena cava, including the double inferior vena cava or isolated left inferior vena cava, are uncommon and of great clinical importance. Inferior vena cava variations signify predisposition to deep vein thrombosis and may complicate retroperitoneal surgeries including abdominal aortic surgery. Failure to recognize such variations may predispose a patient to life-threatening complications. This prospective anatomical study assessed 129 cadavers for variations of the inferior vena cava. One of the 129 cadavers (0.78%) possessed a double inferior vena cava and none (0%) possessed an isolated left inferior vena cava. The left-sided inferior vena cava was of a larger diameter than that of the right-sided inferior vena cava — opposite of what would be seen in a Type III duplication. Therefore, this observation expands the three-type classification system to include a Type IV duplication.

Keywords: anatomy, anatomical variation, deep vein thrombosis, inferior vena cava.

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Introduction

Abnormal development of the inferior vena cava (IVC) has been the subject of anatomical study for centuries [1–5]. In modern times, the study of IVC variation remains in the scope of the anatomical interest, especially in light of advances in the wide variety of surgeries performed in the abdominopelvic region.

During early development, the infrahepatic inferior vena cava (IVC) arises from a set of three pairs of parallel veins between four and eight weeks of life — the posterior cardinal, subcardinal, and supracardinal veins [6–8]. The derailment in the formation of anastomoses, regression of structures that should persist and, conversely, persistence of structures that should regress, leads to variations of the IVC which include the duplicated inferior vena cava (DIVC) and isolated left inferior vena cava (ILIVC). Variations of the IVC signify a predisposition to deep vein thrombosis [9, 10]. When present, IVC variations may occur with concomitant variations including a retrocaval ureter [11–12]. With regard to imaging, IVC variations often masquerade as other pathologies, thereby causing confusion with diagnosis and management [10, 13, 14]. Failure to recognize such variations may lead to life-threatening complications [15–16].

A recent meta-analysis assessing the prevalence of IVC duplication documented only three reports that have assessed the duplication of the IVC via autopsy in the 21st century [17]. In addition to the paucity of modern autopsy study, the aforementioned studies included only populations from Thailand, Egypt, and Poland [17]; therefore, the detailed study of IVC variation via autopsy has also been limited geographically. Furthermore, detailed knowledge of IVC variations by direct observation are of importance with regard to the advancement of the numerous operative techniques performed in the abdominal region. Accordingly, this study aims to assess the prevalence, morphology, and morphometry of IVC variations by direct observation among a modern cadaveric sample in the United States of America.

Materials and Methods

This study was conducted with approval of the West Virginia Anatomical Board. The study design was that of a prospective assessment of IVC variations among cadavers. Cadavers were screened for the DIVC and ILIVC. Altogether a total of 129 adult human cadavers (64 white females and 65 white males) were screened. The average age at death of the sample was 79.2 ± 11.0 years (Mean \pm SD) (Females: 79.3 ± 11.8 years; Males: 79.2 ± 10.1 years). Hence, the population was conceived and subsequently underwent embryological development in the approximate timeframe of the years 1930 to 1950.

When variation was encountered, digital photographs were taken (Canon PowerShot SX50 HS, 12.1 Megapixel). Also, digital calipers (Mitutoyo 0–8 in (0–203.2 mm) ABSOLUTE™ digimatic caliper series 500, accuracy ± 0.025 mm) were included in the frame of the photos as a fiducial for photogrammetry which was subsequently performed with ImageJ software [18, 19].

Results

Variations included one DIVC (1:129; 0.78%) and zero ILIVC (0:129; 0%). The DIVC was found in a 66-year-old white male, whose death resulted from an aortic dissection (Fig. 1). A right-sided IVC ($\varnothing = 16$ mm), and a left-sided IVC ($\varnothing = 26$ mm) flanked an aorta that possessed a large aneurysm ($\varnothing = 58$ mm) that was previously managed with an endovascular stent graft. There was also enlargement of the thoracic aorta, similarly managed with a stent graft. The right IVC drained the right gonadal (testicular) vein ($\varnothing = 7$ mm). Contributing to the left IVC was the left gonadal vein, left renal vein, and left suprarenal vein ($\varnothing = 5, 17,$ and 5 mm, respectively). The left gonadal and renal veins joined the left IVC at the same location. The left IVC traversed the midline, anterior to the aorta to join with the right IVC at an 85° angle. The right renal vein ($\varnothing = 15$ mm), right IVC, and left IVC joined together to form a common IVC ($\varnothing = 32$ mm). The right suprarenal vein ($\varnothing = 3$ mm) drained to posterior aspect of the right renal vein. The right kidney presented an exophytic space-occupying lesion on its anterior surface that was dark in color, nodular, and solid. The tumor measured 21 mm \times 19 mm.

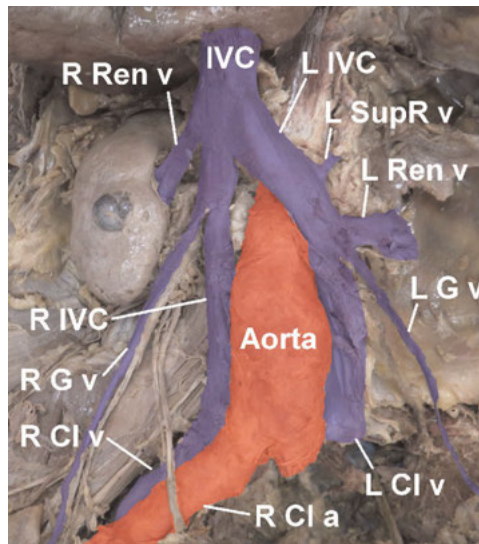


Fig. 1. Anterior view of the abdominal vasculature revealing a duplicated inferior vena cava. Aorta with an infrarenal abdominal aortic aneurysm; L CI v — left common iliac vein; L G v — left gonadal (testicular) vein; L IVC — left inferior vena cava; L Ren v — left renal vein; L SupR v — left suprarenal vein; R CI a — right common iliac artery with aneurysm; R CI v — right common iliac vein; R G v — right gonadal (testicular) vein; R IVC — right inferior vena cava; R Ren v — right renal vein.

Discussion

In the light of the morphological and embryological studies [3, 4, 20–24] duplication of the IVC can be classified into three types termed as:

- Major duplication (Type I) — bilaterally symmetrical trunks and a preaortic trunk of approximately the same size.
- Minor duplication (Type II) — bilaterally symmetrical trunks of approximately the same size but smaller than the preaortic trunk.
- Asymmetric duplication (Type III) — disproportion between left and right IVC (left IVC that is smaller in diameter than the right IVC) and variable size of the preaortic trunk.

The variation detailed in this report did not fit into any of the aforementioned classification types and, therefore, this report describes a novel Type IV variation wherein the left IVC is larger in diameter than the right IVC.

When there is persistence of both the left and right supracardinal veins during development, two distinct IVC result [7, 21]. The left IVC would then typically end at the level of the left renal vein, crossing the midline to join the right IVC [6].

Other embryological derailment has been reported alongside the presence of a DIVC. For example, some have reported interiliac veins occurring in conjunction with a DIVC [25–29]. Also, the DIVC has also presented with a concurrent right-sided preaortic iliac confluence, the so-called “marsupial cava” [30]. The marsupial cava variant was not seen in this case.

Other concomitant variations with embryological etiology have included a congenital vitelline band, which was implicated in the intestinal obstruction in an adult with a DIVC [31]. Further, ureter anomalies, including a retrocaval ureter, otherwise known as a circumcaval ureter, have also been reported to occur in conjunction with the DIVC [11, 12].

The DIVC can complicate surgery for abdominal aortic aneurysm (AAA) [10, 16, 32]. The man reported in this study had an AAA ($\varnothing = 58$ mm) that was previously managed with an endovascular stent graft. This report documents the DIVC occurring alongside an AAA — a pathology in which the surgical management is complicated by the presence of a DIVC. Therefore, it is noteworthy that the individual detailed in this report died as a result of an aortic dissection. It is also important to note that, while, if unrecognized, a DIVC may predispose the patient to serious life-threatening bleeding complications during aortic surgery, the surgery has been performed with favorable results [16].

In addition to considerations regarding the DIVC and abdominal aortic surgery, the presence of the DIVC is important in avoiding adverse events during retroperitoneal surgeries, in general [10]. For example, the DIVC may cause complications with kidney harvesting [33, 34]. Though, several reports have documented successful

nephrectomy in the presence of DIVC [35–37]. The accurate diagnosis of a DIVC is important. The misdiagnosis of a DIVC has caused surgical confusion between a left IVC, which merged with the left renal vein, and the left gonadal vein, leading to the severing of the left IVC during radical nephroureterectomy [15]. The DIVC may be mistaken as para-aortic lymphadenopathy on CT [10, 13, 14]. Also, venous aneurysms have also been reported with a DIVC a scenario which may also cause confusion during imaging interpretation [38].

The DIVC, along with other IVC malformations, are associated with approximately 5% of deep vein thrombosis cases [9, 10]. However, IVC filters have been successfully placed and removed in duplicated IVCs [26, 39–42]. In cases of recurrent pulmonary embolisms after the placement of an IVC filter, screening for a DIVC should be performed. If a DIVC is discovered during the screening, options may include placing filters in both IVC, in the common suprarenal IVC, or performing coil embolization of the smaller ICV [7, 26, 39, 40, 42, 43].

Other procedures involving a DIVC have also been performed successfully. For example, transcaval transcatheter aortic valve replacement have been performed despite DIVC [44]. In the case of a DIVC, a right IVC with leiomyosarcoma has been completely resected [9]. Further, nutcracker syndrome has been reported in conjunction with a DIVC; successfully treated with stent placement and anticoagulants [45].

Conclusions

Inferior vena cava variations arise from embryologic derailment of precursor veins. Such variations include a DIVC and ILIVC. This report identified a DIVC prevalence of 0.78% and an ILIVC prevalence of 0%; therefore, the DIVC and ILIVC should be considered rare findings. This report detailed a DIVC that occurred with a concomitant AAA. Moreover, the variation was that of a novel type IV duplication — a particularly rare finding that warrants additional consideration.

Conflict of interest

The authors have no conflicts of interest to report. The authors have no financial contributions to disclose.

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