Complete Duplication of Inferior Vena Cava Coexisting with Double Superior Vena Cava In Situ Solitus: Hitherto Unreported Pattern

Arun Sharma[1] Harsimran Bhatia[1,*] Sanjeev Hanumantacharya Naganur[2] Manphool Singhal[1]

[1] Department of Radiodiagnosis, Postgraduate Institute of Medical Education and Research, Chandigarh, India
[2] Department of Cardiology, Postgraduate Institute of Medical Education and Research, Chandigarh, India

Address for correspondence Arun Sharma, DM, Department of Radiodiagnosis, Postgraduate Institute of Medical Education and Research, Chandigarh 160012, India (e-mail: drarungautam@gmail.com).

Indian J Radiol Imaging 2024;34:177–180.

Abstract

Congenital anomalies of inferior vena cava are increasingly being recognized with the technical advancements and increased utilization of cross-sectional imaging techniques. Duplication of inferior vena cava classically involves duplication of the infrarenal segment, where both inferior vena cava ascend on either side of the abdominal aorta until they form a confluence at the level of the renal veins. It has been extensively described in literature with few reports of more complex variation in the form of duplicated infrarenal inferior vena cava with azygos or hemiazygos continuation. This article describes extremely rare complete duplication of inferior vena cava involving both suprarenal and infrarenal segments. Moreover, the complete duplication of inferior vena cava is seen in association with concomitant double superior vena cava, in a patient with visceroatrial situs solitus and associated congenital heart disease, which to the best of our knowledge, has not been reported so far in literature. This study also highlights the utility of multidetector computed tomography in accurate identification of such anomalies.

Keywords

- IVC duplication
- complete duplication of IVC
- double IVC
- double SVC
- IVC interruption
- azygos or hemiazygos continuation
- SVC anomalies
- IVC anomalies

Introduction

Inferior vena cava (IVC) anomalies result from aberrations in the complex, multistep, and multisegmental embryological development process, depending on persistence and/or regression of components of the vitelline and the cardinal venous system. They may be isolated or seen associated with situs abnormalities and congenital heart diseases.[1,2] Although double IVC has also been reported with azygos or hemiazygos continuation, complete duplication of IVC involving both suprarenal and infrarenal segments is an extremely rare entity with only a few cases reported so far. Moreover, to the best of our knowledge, complete duplication of IVC in association with concomitant duplicated superior vena cava (SVC) has not been reported so far. We describe this rare systemic venous pattern in a 6-month-old infant with situs solitus and associated congenital heart disease.

Case Presentation

Computed tomography (CT) angiography (performed on 2 × 192-slice dual source CT [Somatom Force, Siemens Medical

* *Contributed equally with the first author and shares the first authorship.*

article published online
October 27, 2023

DOI https://doi.org/10.1055/s-0043-1775796.
ISSN 0971-3026.

Thieme Medical and Scientific Publishers Pvt. Ltd., A-12, 2nd Floor, Sector 2, Noida-201301 UP, India

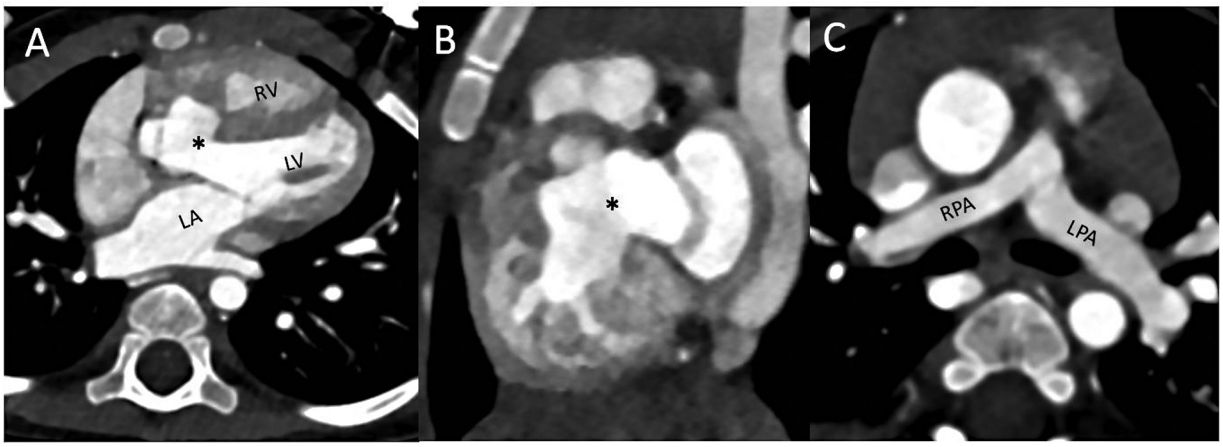

Fig. 1 Computed tomography angiography images (**A–C**) showing features of tetralogy of Fallot, including subaortic ventricular septal defect (*) with aortic override, infundibular–valvular pulmonary stenosis and right ventricular hypertrophy with confluent pulmonary arteries. LA, left atrium; LPA, left pulmonary artery; LV, left ventricle; RPA, right pulmonary artery; RV, right ventricle.

Solutions, Forchheim, Germany] using nonionic iodinated contrast [1.5 mL/kg at the rate of 1.5 mL/second with 10–12 mL of saline chase] with dual phase [initial gated followed by nongated flash] acquisition) from a 6-month-old infant with cyanosis showed situs solitus, levocardia with features of tetralogy of Fallot, including subaortic ventricular septal defect with aortic override, infundibular and valvular pulmonary stenosis, and right ventricular hypertrophy (►**Fig. 1**). Pulmonary arteries were confluent and good sized. Small ductus arteriosus was seen with occluded pulmonary end. Aortic arch was left sided with normal branching pattern. Pulmonary venous drainage was normal with distinct anomalous pattern of systemic venous drainage. Double SVC was seen with no intercommunicating vein. Right SVC was seen draining into right atrium with left SVC draining into the right atrium via coronary sinus. The azygos and hemiazygos veins were dilated. There was complete duplication of IVC with the presence of two venous channels (right larger than left) along both sides of

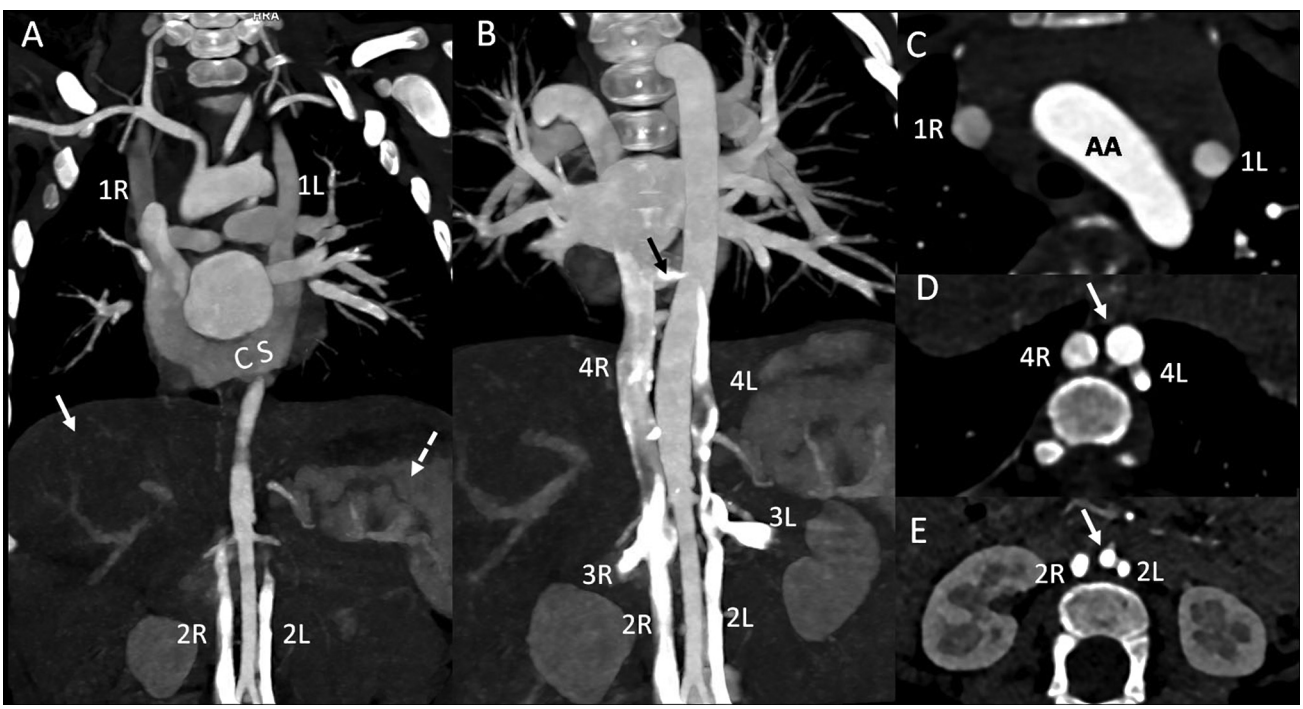

Fig. 2 Computed tomography angiography images; (**A–B**) Maximum intensity projection coronal images. (**A**) Double superior vena cava (SVC); right-sided SVC (1R) draining into right atrium and left-sided SVC (1L) draining into right atrium via coronary sinus (CS). (**B**) Double inferior vena cava (IVC); right IVC (2R) and left IVC (2L) draining right renal vein (3R) and left renal vein (3L) and continuing as azygos vein (4R) and hemiazygos vein (4L), respectively; black arrow denotes drainage of hemiazygos vein into azygos vein. Liver (white arrow in A), spleen (dashed white arrow in A), gastric fundus and systemic venous atrium are seen in normal anatomical positions. (C–E): Axial images (superior to inferior sections). (**C**) Double SVC (1R: right SVC and 1L: left SVC) on both sides of the aortic arch (AA); (**D**) Suprarenal continuation of duplicated IVC 4R: Azygos continuation and 4L: Hemiazygos continuation along both sides of abdominal aorta (white arrow); (**E**) Infrarenal duplication of IVC: (2R: right IVC and 2L: left IVC) along both sides of abdominal aorta (white arrow).

the abdominal aorta. Right-sided IVC received the right renal vein and continued as azygos vein with IVC interruption. Left-sided IVC received the ipsilateral renal vein and continued as hemiazygos vein that courses posterior to thoracic aorta to drain into azygos vein at D9 vertebral level. Azygos vein was seen draining into right SVC (►**Figs. 2**–**3**). The right and middle hepatic veins were not opacified on CT. Left hepatic vein was seen draining separately into the right atrium near the site of coronary sinus drainage. Ultrasound correlation showed drainage of right and middle hepatic veins into right atrium via small suprahepatic IVC. Coronary arteries were normal. No significant aortopulmonary collaterals were seen. No airway or lung parenchymal abnormality was seen.

Discussion

Duplication of IVC or commonly referred as double IVC is seen in approximately 0.2 to 3% of the population and typically comprises of two infrarenal IVCs.[3] Persistence of both the supra-cardinal veins results in this prototypical type of duplication. In the commonest form, both IVCs ascend on either side of the abdominal aorta with left-sided IVC crossing anterior to abdominal aorta (preaortic trunk) at the level of renal veins to join the right IVC.[4] The left IVC may also drain into left renal vein rather than draining directly into right IVC.[5] In the less common form, both the infrarenal IVCs lie on the same side, most often on the right side.[6] Recurrent pulmonary embolism despite placement of an IVC filter may give a clinical hint to the presence of this anomaly. Variations also exist in the course and the site of drainage of double IVC, resulting in several patterns. Double IVC with hemiazygos continuation may be seen with right-sided IVC draining into a retroaortic right renal vein. Similarly, left-sided IVC may drain into a left-sided retroaortic renal vein and further continue as azygos vein.[7] All of these, however, consist of duplication of only the infrarenal segments terminating below the hepatic level with distal drainage via the azygos or hemiazygos systems.

Complete duplication of IVC involving both the infrarenal and suprarenal segments is extremely rare. Literature search showed two rare cases of complete duplication of IVC where continuous azygos–hemiazygos veins joined together above the diaphragm and drained into SVC in one case.[8] In the second case, right IVC followed the normal course and drained into the right atrium, whereas the left IVC continued as hemiazygos vein and drained into SVC.[9] Our case is rare in demonstrating the complete IVC duplication incorporating both infrarenal and suprarenal segments with continuous azygos–hemiazygos veins that joined supradiaphragmatically and drained into right SVC in the presence of double SVC with left SVC draining into right atrium via coronary sinus. No such case has been described previously with coexisting double SVC with complete duplication of IVC.

Double SVC with persistent left SVC is often incidentally detected in approximately 0.3% of general population and 10 to 11% of patients with congenital heart disease and results from failure of left precardinal and common cardinal vein to atrophy.[10,11] While heterotaxy syndromes are seen in nearly half of such cases, other coexisting cardiac anomalies range from atrioventricular septal defect to tetralogy of Fallot and single ventricle.[1,12] Although left SVC in such cases usually drains into right atrium via coronary sinus, drainage into left atrium causing significant hemodynamic alteration is seen in approximately 10 to 20% of the cases.[13]

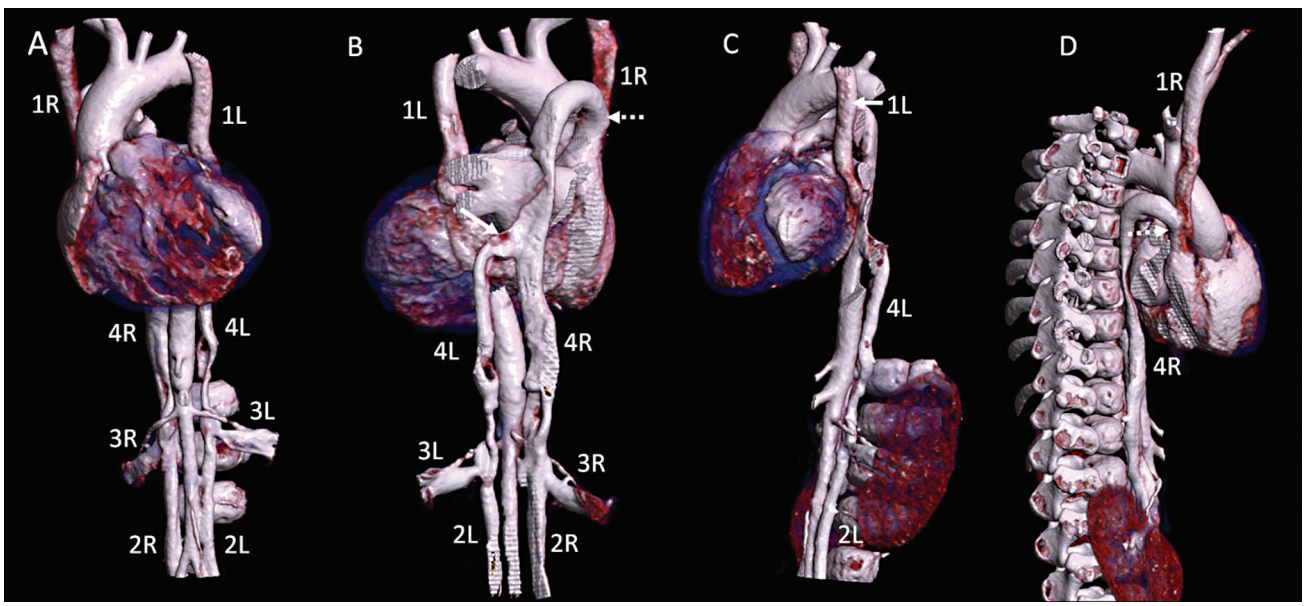

Fig. 3 Volume-rendered computed tomography images; (**A**) Anterior view, (**B**) posterior view, (**C**) right lateral view, and (**D**) left lateral view demonstrating double superior vena cava (SVC) with complete inferior vena cava (IVC) duplication. 1R: Right SVC, 1L: left SVC, 2R: right IVC, 2L: left IVC, 3R: right renal vein, 3L: left renal vein, 4R: suprarenal azygos continuation, 4L: suprarenal hemiazygos continuation. White arrow in B denotes drainage of hemiazygos vein into azygos vein. White arrow in C denotes left SVC (1L). Dashed white arrows in B and D denote azygos vein drainage into right SVC.

Accurate identification of anomalous venous channels on cross-sectional imaging is essential as a part of preoperative evaluation, in planning catheter insertion, deciding cannulation strategies, and accurate mapping for intracardiac repair.[5,13,14] The course, caliber, and drainage points of such channels must be clearly delineated and communicated to the treating surgeon and the anesthesiologist. Multidetector CT allows accurate identification of systemic venous anomalies in addition to delineation of associated conditions, thereby helping in proper surgical planning.

Funding
None.

Conflict of Interest
None declared.

References

1 Lendzian T, Vogt J, Krasemann T. Are anomalies of the caval veins more common in complex congenital heart disease? Herz 2007; 32(08):657–664
2 Spentzouris G, Zandian A, Cesmebasi A, et al. The clinical anatomy of the inferior vena cava: a review of common congenital anomalies and considerations for clinicians. Clin Anat 2014;27(08):1234–1243
3 Phillips E. Embryology, normal anatomy, and anomalies. In: Ferris EJ, Hipona FA, Kahn PC, Phillips E, Shapiro JH, eds. Venography of the Inferior Vena Cava and Its Branches. Baltimore, MD: Williams & Wilkins; 1969:1–32
4 Natsis K, Apostolidis S, Noussios G, Papathanasiou E, Kyriazidou A, Vyzas V. Duplication of the inferior vena cava: anatomy, embryology and classification proposal. Anat Sci Int 2010;85 (01):56–60
5 Ang WC, Doyle T, Stringer MD. Left-sided and duplicate inferior vena cava: a case series and review. Clin Anat 2013;26(08): 990–1001
6 Nagashima T, Lee J, Andoh K, et al. Right double inferior vena cava: report of 5 cases and literature review. J Comput Assist Tomogr 2006;30(04):642–645
7 Bass JE, Redwine MD, Kramer LA, Huynh PT, Harris JH Jr. Spectrum of congenital anomalies of the inferior vena cava: cross-sectional imaging findings. Radiographics 2000;20(03):639–652
8 Petik B. Inferior vena cava anomalies and variations: imaging and rare clinical findings. Insights Imaging 2015;6(06):631–639
9 Shaheen S, Alyahya KI, El Fouhil AF, et al. An extremely rare complete bilateral duplication of inferior vena cava in a male cadaver: anatomy, embryology and clinical relevance. Folia Morphol (Warsz) 2022;81(01):247–253
10 Buirski G, Jordan SC, Joffe HS, Wilde P. Superior vena caval abnormalities: their occurrence rate, associated cardiac abnormalities and angiographic classification in a paediatric population with congenital heart disease. Clin Radiol 1986;37(02):131–138
11 Gris P, Wilmet B, Benchillal A, et al. [Persistent left superior vena cava. Apropos of 2 cases]. Rev Pneumol Clin 1995;51(01):33–35
12 Nagasawa H, Kuwabara N, Goto H, et al. Incidence of persistent left superior vena cava in the normal population and in patients with congenital heart diseases detected using echocardiography. Pediatr Cardiol 2018;39(03):484–490
13 Azizova A, Onder O, Arslan S, Ardali S, Hazirolan T. Persistent left superior vena cava: clinical importance and differential diagnoses. Insights Imaging 2020;11(01):110
14 Sitwala PS, Ladia VM, Brahmbhatt PB, Jain V, Bajaj K. Inferior vena cava anomaly: a risk for deep vein thrombosis. N Am J Med Sci 2014;6(11):601–603

