CT presentations of anatomic variants in superior vena cava system

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Learning objectives

The purpose of this paper was to overview various CT presentations of different venous anatomic variants in the system of superior vena cava (SVC) in association with anatomic variants of azygos veins.
Background

Formation of three great venous systems: cardinal, supracardinal and subcardinal occurs during embryonic period. Right and left superior vena cava are formed predominantly from anterior cardinal veins. Azygos and hemiazygos veins are formed from anterior parts of supracardinal veins. Left parts of both systems merge to form proximal part of coronary sinus.

Origin of right and left inferior vena cava is complex. Partially, they are derived from posterior parts of supracardinal veins - this explains possibility for continuation of right and left inferior vena cava into azygos and hemiazygos vein. ¹)

Persistent left superior vena cava is a congenital venous anomaly occurring in approximately 0.3% to 0.5% of the normal population²) and in that form it is the most common congenital anomaly of the systemic veins. Also, it is often associated in patients with other documented congenital heart abnormalities - up to 12%. ³)

Hemiazygos involvement as continuation of the left inferior vena cava into hemiazygos vein or continuation of left superior vena cava into accessory hemiazygos vein is extremely rare.
Fig. 1: During the development cardinal veins (future right and left superior vena cava) and supracardinal veins (future azygos veins) are separated. They merge from the left to the right side postpartally. (VCS - superior vena cava; VBC - brachiocephalic vein)

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Persistent left superior vena cava (LSVC) is usually completely separate from the right superior vena cava with agenesis of the left brachiocephalic vein. In cases in which left brachiocephalic vein is present, it can be either dominant or hypoplastic and that determines the presentation of persistent LSVC which can also be either dominant or hypoplastic.\textsuperscript{4)}

Fig. 2: Schematic and CT presentation of dominant left superior vena cava with hypoplastic left brachiocephalic vein. (VCS - superior vena cava; VBC - brachiocephalic vein)

References: KBC Zemun - Zemun/RS

Persistent LSVC forms during thoracic development due to the failure of obliteration of left anterior cardinal vein. When left anterior cardinal vein remains completely patent, left brachiocephalic vein will not develop.\textsuperscript{5)} If LSVC obliteration process is only partially achieved, then different form of venous presentation will persist postpartally, either dominant left brachiocephalic vein or dominant LSVC.\textsuperscript{4)}
Fig. 5: Schematic and CT presentation of hypoplastic left superior vena cava draining in coronary sinus (common variant) with dominant left brachiocephalic vein. (VCS - superior vena cava; VBC - brachiocephalic vein)

References: KBC Zemun - Zemun/RS Very rarely LSVC can continue to accessory hemiazygous vein without anastomosing with coronary sinus and then ending in right superior vena cava. Other smaller veins can also merge with LSVC and then become more prominent in diameter, such as left pericardiacophrenic vein in our example. Left brachiocephalic vein is present and it is of dominant type, as well as hypoplastic LSVC which drains into right superior vena cava just above the junction of azygos vein. 6)
Fig. 7: Schematic and CT presentation of atypical hypoplastic left superior vena cava joining right superior vena cava just above the junction of azygos vein. (VCS - superior vena cava; VBC - brachiocephalic vein)

**References:** KBC Zemun - Zemun/RS

Hemiazygos vein can be dominant to the azygos vein when there is anomalous continuation of persistent left inferior vena cava into hemiazygos vein.\(^7\) Size of left inferior vena cava is dependent of the amount of blood volume from left common iliac vein that drains in it, and is inversely correlated to the amount of blood volume that inflow in right inferior vena cava from left common iliac vein.

![Figure 7: Schematic and CT presentation of atypical hypoplastic left superior vena cava joining right superior vena cava just above the junction of azygos vein. (VCS - superior vena cava; VBC - brachiocephalic vein).](image)

Fig. 10: Schematic and CT presentation of hypoplastic left inferior vena cava (LIVC). There is a dominant left iliac arch draining into right inferior vena cava and smaller LIVC which joins left renal vein to form hemiazygos vein. Hemiazygos vein merges with azygos vein and then blood from LIVC ends up in superior vena cava. (VCS - superior vena cava; VCI - inferior vena cava).

**References:** KBC Zemun - Zemun/RS
Fig. 3: Typical radiographic presentation of the LSVC with the widening of upper mediastinum to the left and comparative CT representing duplication of superior vena cava. (VCS - superior vena cava)

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Fig. 4: CT presentation of separate endings of azygos and hemiazygos veins into right and left superior vena cava respectively. Both right and left vena cava are of the same diameter when there is a combination of dominant left VCS and hypoplastic left VBC. (VCS - superior vena cava; VBC - brachiocephalic vein)

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**Fig. 6:** CT presentation of hypoplastic LSVC (proximal and distal half) with common ending in coronary sinus and right atrium. (VCS - superior vena cava)

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**Fig. 8:** CT presentation for proximal half of the atypical LSVC. After junction with left pericardiacobphrenic vein it continues to the opposite-right side. (VCS - superior vena cava)

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Fig. 9: CT presentation for distal half of the atypical LSVC that goes in right superior vena cava along the route of accessory hemiazygos vein, just above the ending of azygos vein. (VCS - superior vena cava)

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Fig. 11: CT presentation of hypoplastic left inferior vena cava (LIVC) that continues as hemiazygos vein. Notice that hemiazygos vein diameter is much greater compared to diameter of juxtaphrenic part of azygos vein due to the presence of LIVC.

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Conclusion

Anatomic variants of superior vena cava system are very diverse and even though they are usually incidental finding, the knowledge about these variants is important for their adequate identification, careful analysis and correct interpretation, especially in pre-procedural diagnostic settings for cardiac catheterization and cardiovascular surgical interventions.
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