A very rare abnormal course of the thoracic venous system: Long journey to the right ventricle

Yao Chang Wang, MBBS, An Ning Feng, MD, Wei Hsian Yin, MD, Mason Shing Young, MD

From the Cardiovascular Division, Department of Internal Medicine, Cheng-Hsin General Hospital, Taipei, Taiwan.

Introduction
Knowledge of the normal and abnormal anatomy of the thoracic venous system is very important for any cardiac electrophysiologist who performs radiofrequency ablation and transvenous pacemaker implantation. In this article, we report on a 68-year-old male subject with a very rare abnormal long course of the thoracic venous system.

Case report
The patient had a history of coronary artery disease post-coronary artery bypass graft and was hospitalized for syncope due to atrial fibrillation with slow ventricular response, and underwent ventricular pacemaker implantation (VVIR mode). Under fluoroscopic guidance, a right ventricular screw-in lead was inserted via the cephalic vein. The course of the pacemaker leads deviated from the normal brachiocephalic pathway and ran dorsally along the left lateral aspect of the thoracic vertebra and across the thoracic vertebra to the right side at T6 level, and then ran to the right atrium via the superior vena cava (SVC), as shown in Figure 1 (A,B). Anomalous communication with the coronary sinus via the superior vena cava (SVC), as shown in Figure 1 (C,D). Knowledge of the normal and abnormal anatomy of the thoracic venous system: Long journey to the right ventricle

Discussion
Embryology of thoracic venous system
Around day 17 of fetal development, blood vessel formation occurs within the splanchnopleuric mesoderm of the yolk sac. Blood islands within the yolk sac may be observed at day 21. Central parts of the islands host hemoblasts and the outer layers form the blood vessels.1-3 A system of 3 paired veins is found in the fourth week of gestation, which includes the umbilical veins from the chorion, the vitelline veins from the yolk sac, and the cardinal veins from the body of the embryo itself, all of which open to the right and left horns of the sinus venosus of the heart. Vitelline veins develop to part of the inferior vena cava (IVC), hepatic veins, hepatic sinusoid, ductus venosus, portal vein and its tributaries. Umbilical veins contribute to the hepatic sinusoids; the left umbilical vein forms ligamentum teres. Cardinal veins develop into anterior, posterior, subcardinal, and supracardinal veins.2,3

Anterior cardinal veins (ACVs) drain cranial parts of the body and posterior cardinal veins (PCVs) provide drainage from caudal parts. ACV and PCV join together into the common cardinal vein (Cuvier ducts), entering the sinus venosus of the early heart. At the eighth week of fetal life the left brachiocephalic vein is being formed, connecting the left and right ACVs. The portion of the left ACV below this connection partially obliterates, forming the “ligament of Marshall.” The distal remaining section of the left ACV forms the coronary sinus and oblique vein of the heart. The right ACV remains patent and, together with the right common cardinal vein, becomes the precursor of the SVC system. Most PCVs undergo atrophy and their patent remnants form the renal segment of the IVC and common iliac veins. Simultaneously, subcardinal and supracardinal veins are being formed that are involved in development of IVC. Supracardinal veins (SV) give the origin to theazygos system of veins. Usually SVs develop anastomosis at the level of the thoracic spine. The right SV becomes the AZV. The left one below anastomosis transforms into the hemiazygos vein (HAZV) and above the anastomosis at the T6 level and finally drained into the right SVC, as shown in Figure 1 (C,D).

KEYWORDS Pacemaker implantation; Thoracic venous system; Azygos vein; Accessory hemiazygos vein; Right ventricle
ABBREVIATIONS ACV = anterior cardinal vein; HAZV = accessory hemiazygos vein; AZV = azygos vein; CT = computed tomographic; HAV = hemiazygos vein; IVC = inferior vena cava; PCV = posterior cardinal vein; SV = supracardinal vein; SVC = superior vena cava

Address reprint requests and correspondence: Yao Chang Wang, MBBS, No. 214, 8F-1, Zhong Hsin St, Zhong He District, New Taipei City, 235, Taiwan. E-mail address: aididminthant@gmail.com.
obliterates. In some cases only the cranial section of the left SV remains patent as AHAZVs.\(^3,4\)

**Anatomy of azygos system**

The azygos system forms a connection between the SVC and IVC and serves as an important collateral pathway when there is obstruction of the IVC or SVC. It is a paired paravertebral venous pathway in the posterior thorax. At T5–T6, the AZV arches ventrally just cephalad to the right main bronchus and drains into the SVC. At approximately T8–T9 the HAZVs join the AZV, usually posterior to the aorta. The accessory hemiazygos joins the azygos at T7–T8, crossing posterior to the aorta.\(^5,6\) Usually, the accessory hemiazygos and hemiazygos form common channels posterior to the aorta. The AHAZV, also called the superior HAZV, drains the superior left hemithorax. In a majority of cases there is a small connection to the left superior intercostal vein, and rarely, 1%–2% of the time, the AHAZV drains into the brachiocephalic vein.\(^6,7\)

**Issue regarding lead extraction and lead reinsertion**

Pacemaker leads that traverse via the AZV are difficult to extract. Three and a half years after follow-up in our patient, the pacemaker function revealed 57.4% sensing and 42.6% pacing, 7 years of battery longevity remained, and the ventricular lead had a threshold of 0.75 volts at 0.4

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**Figure 1** Abnormal course of thoracic venous system. **A:** Anteroposterior and **B:** lateral views of chest radiograph revealed a pacemaker lead that traveled via abnormal route to the right ventricle. **C, D:** Abnormal course of thoracic venous system confirmed by electrocardiographic-gated 64-slice computed tomographic angiography with a 3-dimensional reconstruction. AHAZ = accessory hemiazygos vein; AZ = azygos vein; SVC = superior vena cava; LBC = left common brachiocephalic vein; RA = right atrium.
milliseconds and impedance of 487 ohms and R wave within the range of 5.6–8 millivolts. If a failure in pacemaker lead function occurs, insertion of the new pacemaker leads via the right side was planned and arranged CT angiography of the right thoracic venous system. Unfortunately, deterioration of the patient’s renal function test occurred during these years, with a recent serum creatinine level of 3.4 mg/dL (progression of diabetic nephropathy). For the sake of the patient, CT angiography was withheld. Revision of previous chest CT scan revealed that the right thoracic venous system seems to be normal.

Conclusion
An aberrant thoracic venous system was found in our patient during implantation of pacemaker leads. The lead ran from a stump of the brachiocephalic vein to the AHAZV, which crosses through the thoracic vertebra posteriorly to the AZV and enters the right atrium via the SVC. Drainage of the accessory hemiazygos to the left brachiocephalic vein is very rare. The knowledge of this anomalous communication enabled the cardiac electrophysiologist to perform the procedure smoothly and without any hesitation.

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