Response to Letter Regarding Article, “Bilateral Superior Venae Cavae With Crisscross Atrial Drainage”

We acknowledge the letter from Murakami and colleagues about our article, “Bilateral Superior Venae Cavae With Crisscross Atrial Drainage.”

In that article, we documented a rare thoracic systemic venous drainage anomaly in which a persistent left superior vena cava was seen draining into the right atrium (RA) via the coronary sinus and the right superior vena cava (RSVC) draining into the left atrium (LA) via the LA orifice of the right superior pulmonary vein (RSPV). We believe that our description of the anomaly is correct not only hemodynamically but also anatomically. Unroofing of the RSPV into the RSVC andatomic drainage of the RSVC into the LA via the LA orifice of the RSPV were confirmed by computed tomography angiography. As discussed in our article and elsewhere, embryologically, this anomaly is thought to be attributable to a deficiency in the posterior wall of the RSVC and the anterior wall of the contiguous RSPV, thus creating an anomalous venous confluence that can accurately be called a cavopulmonary venous defect, not just a sinus venous defect, because the sinus venous part (the smooth posteromedial wall) of the RA is not part of the confluence. Its anatomic variations include the sinus venous defect of the SVC (or high) type and the bialtral and the left atrial drainage of the RSVC. This cavopulmonary venous defect leads to unroofing of the RSPV into the RSVC, and the LA orifice of the RSPV becomes the interatrial communication, but it cannot be regarded as a defect in the interatrial septum (septum primum or septum secondum). Predominance of this interatrial communication in fetal life leads to diminution of flow through the RA orifice of the SVC and consequent RSVC orifice atresia.

The room air systemic oxygen saturation of our patient was nearly normal, ≥96%, which was well above the systemic desaturation causing visible central cyanosis (<79% in nonanemic individuals). Similarly, our patient did not have any other symptoms of significant right-to-left shunt such as effort intolerance. A possible explanation for the absence of systemic desaturation and significant right-to-left shunt was the near-complete drainage of the systemic venous return into the RA. All of the inferior vena cava and persistent left superior vena cava blood was draining into the RA and getting normally oxygenated. Furthermore, on the axial computed tomography images, the accessory hemiazygous vein (draining into the persistent left superior vena cava) was almost the same size as or was slightly bigger than the ayzygous vein (draining into the RSPV). Hence, only part of the RSPV blood was shunted from right to left via the cavopulmonary venous defect, which alone was insufficient to cause systemic desaturation and visible cyanosis.

Bilateral SVC with one that is partially or completely atretic can be seen in visceral heterotaxy and right isomerism with transverse liver, asplenia, bilateral morphological right bronchi, bilateral morphological right atrial appendage, bilateral trilobed lung, and bilateral sinoatrial node. However, none of the features of visceral heterotaxy mentioned above were present in our case.

To summarize, by presenting this case, we aimed to make readers aware of this rare and unexpected thoracic systemic venous anomaly that may be encountered during procedures that require upper-limb venous access such as transvenous pacemaker lead implantation and the techniques to overcome this problem. In addition, there would be a need to exercise caution while handling peripheral intravenous cannulas in the right upper limb because LA drainage of the R SVC would make these patients vulnerable for paradoxical systemic embolism.

Disclosures

None.

References

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_Circulation_. 2016;133:e614
doi: 10.1161/CIRCULATIONAHA.116.022198

_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/133/17/e614

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