A neonate was noted to have respiratory distress and progressive cyanosis since birth. A cardiac sonogram of the subcostal 4-chamber view disclosed complex heart disease, including common atrium, dextrocardia, right-sided aortic arch, and single ventricle (Figure 1). An unusually dilated right superior vena cava was noted. Abdominal sonography revealed asplenia with transverse liver. The pulmonary veins, however, were not well depicted due to poor acoustic window. Non-EKG gating multidetector computed tomography (Aquilion, Toshiba) using automatic tube current modulation was performed with 64×0.5-mm slice thickness and contrast enhancement to rule out pulmonary venous anomaly. Three-dimensional reconstructions using a volume-rendering technique with posterior view showed all 4 pulmonary veins drained to a tortuous dilated vertical vein and then coursed into the medial aspect of superior vena cava (Figure 2). A right oblique anterior view with 3D rendering technique showed a giant right superior vena cava resulting from the vertical vein impingement (Figure 3). Junction stenosis at the confluence elucidated the vertical vein dilatation (Figure 4).

Right isomerism (asplenia, heterotaxy syndrome) is usually associated with complex cardiac malformations, including common atrioventricular valve, single ventricle, abnormal ventriculoarterial connections, pulmonary outflow tract obstruction, and total anomalous pulmonary venous connection. The identification of obstructive pulmonary venous return is crucial because of the preoperative planning and prognosis. Several techniques have been proposed for repair of total anomalous pulmonary venous connection, but the mortality remains high for complex cases. Echocardiography is still the first-line imaging modality for congenital heart disease due to freedom from radiation and widespread availability. Meanwhile, multidetector computer tomography with multiplanar and 3D volume reconstruction could serve as a complementary tool because of its excellence in depicting congenital heart anomalies, especially for extracardiac portions.

Disclosures

None.

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Figure 2. DA indicates descending aorta; SVC, superior vena cava; RPA, right pulmonary artery; LPA, left pulmonary artery; VV, vertical vein; PV, pulmonary vein.

Figure 3. DA indicates descending aorta; SVC, superior vena cava; RPA, right pulmonary artery; VV, vertical vein.

Figure 4. Multiplanar coronal computed tomography reconstruction demonstrated the junction stenosis at the confluence (black arrow), single ventricle with dominant left ventricle, and transverse liver. AA indicates aortic arch; SVC, superior vena cava; SVLV, single ventricle with dominant left ventricle; RRV, rudimentary right ventricle; MPA, main pulmonary artery; VV, vertical vein.
A Giant Superior Venous Cava Resulted From Obstructive Total Anomalous Pulmonary Venous Connection
Yi-Shan Tsai, Chang-Hsien Yu, Kwok-Kuen Pang and Shin-Lin Shih

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