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## Multidetector Computed Tomographic Angiography Imaging of Congenital Pulmonary Venous Anomalies: A Pictorial Review

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### Abstract

Congenital pulmonary venous anomalies are not uncommon that can occur either in isolation or in association with different forms of congenital heart disease. Clinical presentation of these anomalies may vary from the relatively benign single anomalous partial pulmonary venous return to life-threatening critical obstructed total anomalous pulmonary venous return. Accurate delineation of these anomalies and accompanied cardiovascular anomalies are crucial to guide decision making in these patients. Low-dose high-pitch dual-source 256-detector multidetector computed tomographic angiography is a fast and reliable imaging modality allowing comprehensive noninvasive anatomic imaging in neonates and children with congenital pulmonary venous anomalies with lower radiation doses and should be preferred for these patients after transthoracic echocardiography.

### Résumé

Les anomalies congénitales des veines pulmonaires sont relativement fréquentes. Elles peuvent se présenter seules ou en association avec d'autres formes de cardiopathies congénitales. Le tableau clinique varie, allant d'un retour veineux pulmonaire anormal partiel (relativement anodin) à des retours veineux pulmonaires anormaux totaux avec obstruction (potentiellement mortels). Il est essentiel de définir ces anomalies, ainsi que les autres anomalies cardiovasculaires qui les accompagnent, en vue de prendre des décisions éclairées à l'égard des patients. L'angiographie par tomographie multibarrettes à 256 détecteurs, faible dose, pas élevé et double source est une modalité d'imagerie rapide et fiable. Elle permet la visualisation complète et non effractive des structures anatomiques des nouveau-nés et des enfants qui présentent des anomalies congénitales du retour veineux pulmonaire, tout en induisant de faibles doses de rayonnement. Il s'agit de la modalité à privilégier chez ces patients après l'échocardiographie transthoracique.

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**Key Words:** Anomalous pulmonary venous return; Dose reduction; Dual-source multidetector computed tomography angiography; Levotriocardinal vein; Malposition of the septum primum; Scimitar syndrome

Congenital pulmonary venous anomalies are frequent in patients with congenital heart disease and they are especially common in patients with atrial and visceral situs anomalies. These anomalies range across the entire spectrum from incidental findings to conditions that are lethal if untreated. Accurate delineation of these anomalies and accompanied cardiovascular anomalies are crucial to guide decision making in these patients. Though congenital pulmonary venous anomalies have been assessed traditionally with echocardiography and catheter angiography, magnetic

resonance (MR) imaging and multidetector computed tomographic (MDCT) angiography are playing increasing roles in diagnosis and follow-up of these anomalies. These imaging modalities help overcome the limitations of echocardiography, including a poor acoustic window and poor depiction of extracardiac vascular structures, as well as limitations of catheter angiography such as overlapping of adjacent vascular structures, difficulty in simultaneously demonstrating systemic and pulmonary vascular systems, catheter-related complications, and relatively high doses of ionizing radiation [1]. Even though the great capabilities of MR imaging for anatomic and functional evaluation of the heart, it is time consuming and may require a lengthy period of patient sedation; therefore, the use of MR imaging in seriously ill or uncooperative patients is often

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limited. In recent times, MDCT angiography has increasingly been used in the assessment of patient with suspected or known congenital heart disease, especially in patients in whom associated vascular anomalies must be ruled out [2]. MDCT scanners with a high volume of coverage (i.e., 128 or more slices per gantry rotation) permit faster and more accurate assessment of the vascular anatomy compared with first-generation multidetector technology, with lower radiation exposure and without the need for sedation [3].

In this article we review embryologic bases and clinical features and we illustrate low-dose dual-source 256-detector MDCT angiography images of congenital pulmonary venous anomalies including total anomalous pulmonary venous return (TAPVR), partial anomalous pulmonary venous return (PAPVR), scimitar syndrome, cor triatrium sinister, levoatriocardinal vein (LACV), congenital pulmonary vein stenosis or atresia, and malposition of the septum primum.

### MDCT Scanning Technique

MDCT angiography examinations were performed using a dual-source 256-MDCT scanner (Definition Flash, Siemens Healthcare, Forchheim, Germany) with a sectional collimation of  $2 \times 128 \times 0.6$ . Non-electrocardiography-gated protocol with pitch factor of 3 is used and every scan was obtained with z-axis modulation technique (CARE Dose; Siemens Healthcare, Erlangen, Germany). The voltage and tube current were adjusted to the patient's weight as follows: 80 kV dosage was used for patients weighing <20 kg, and 100 kV for 20-80 kg, and tube current was 10 mA/kg for patients weighing less than 9 kg, and 5 mA for each additional kg. The FOV was extended from base of the neck to the diaphragm. The imaging data was acquired during an intravenous injection of 1-1.5 mL/kg iodinated contrast agent (Iodixanol, Visipaque 320 mgI/mL; GE Healthcare,

Milwaukee, WI) at a rate of 1-3 mL/s for infants and older children but manually administered in neonatas or infants younger than 1 year. Contrast material was followed with a chaser of 4-15 mL saline solution. The scanning delay is determined with a bolus tracking technique, with control images positioned in the left ventricle in the axial plane. As contrast material was clearly visualized within left ventricular cavity, the scan was initiated with a 2-second delay. In older children, the scan was started 7 seconds after the attenuation of region of interest positioned in the ascending aorta reached 150 Hounsfield units. Images were reconstructed to 1 mm in thickness and interval reconstruction with a 25f kernel filter and were processed on a separate workstation (Syngo.via; Siemens Healthcare, Forchheim, Germany) with multiplanar reformatting, maximum intensity projection, and volume rendering. In this study, the mean age of the patients with pulmonary venous abnormalities was 4.3 years (range 4 days to 15.7 years). The overall mean effective radiation dose was 1.03 mSv (range 0.15-4.25 mSv), and it was 0.52 mSv (range 0.12-0.74) in the patients younger than 1 year old.

### Embryology of the Pulmonary Veins

During the 2 months in embryonic life, the primordial lung buds develop from the foregut and initially drain into splanchnic plexus, which communicates with primordial systemic veins (paired cardinal veins and umbilicovitelline veins). At the same time, an outpouching from the dorsal wall of the primitive left atrium forms a common pulmonary vein. With time, the common pulmonary vein communicates with the portion of splanchnic plexus that drains blood from the lungs. Later, when the lung buds fuse the common pulmonary vein, the connections between the pulmonary veins and the cardinal and umbilicovitelline veins are involuted.

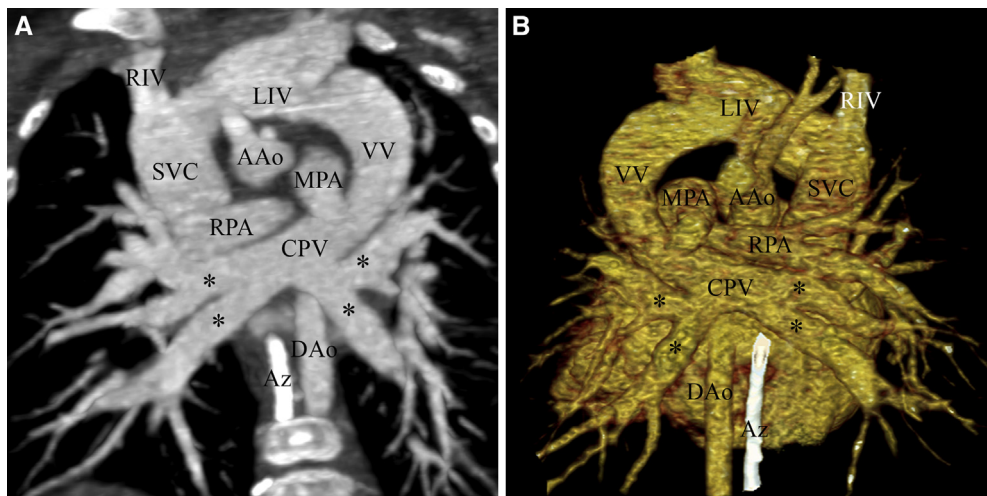


Figure 1. A 3-month-old girl with supracardiac total anomalous pulmonary venous connection to the left innominate vein (LIV). Oblique coronal thin maximum intensity projection (A) and posterior volume rendering (B) images show all pulmonary veins (asterisk) draining into common pulmonary vein (CPV) that connects with LIV via a left-sided ascending vertical vein (VV). LIV and superior vena cava (SVC) are dilated. AAo = ascending aorta; Az = azygos vein, DAc = descending aorta; MPA = main pulmonary artery; RIV = right innominate vein; RPA = right pulmonary artery.

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