

Bland-White-Garland syndrome on coronary CT angiography

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DESCRIPTION

A 2-month-old infant presented with poor weight gain, feeding intolerance and cardiomegaly on chest radiography. Transthoracic echocardiography showed severe left ventricular dilatation and dysfunction, left atrial dilatation, moderate mitral regurgitation and doubtful origin of left coronary artery (LCA). Cardiac catheterisation did not identify the origin of LCA from the aorta. Coronary CT angiography (128-multidetector CT, retrospective ECG-gated with restrictive dose modulation, 70 kVs, 1.2mSv) revealed the origin of the LCA from the pulmonary artery trunk/root junction, immediately above the sinus of Valsalva and was consistent with remaining echocardiography findings (figure 1). The right coronary artery had normal origin. After surgical reimplantation of the LCA in the ascending aorta there was good clinical evolution, with significant weight gain and gradual left ventricular function improvement.

Anomalous LCA from the pulmonary artery, also known as ALCAPA or Bland-White-Garland (BWG) syndrome, is very rare, corresponding to

Learning points

- ▶ Anomalous left coronary artery from the pulmonary artery is very rare and associated with high mortality rate in the first year of life.
- ▶ Timely diagnosis and surgical correction of this condition are crucial for the recovery of myocardial function. Multimodality cardiovascular imaging has a very important role in this setting.
- ▶ Coronary CT angiography allows a non-invasive accurate diagnosis, depicting the origin and course of the coronary arteries. Additionally, it offers a three-dimensional assessment of the anatomic relations between coronary arteries and adjacent structures, useful for surgical planning.

0.25%–0.5% of all congenital cardiac anomalies.^{1,2} BWG syndrome is usually diagnosed in the first 2 months of life and can be isolated or associated with other congenital anomalies.³ There are two types of BWG syndrome: the infant type and the adult type. Without corrective surgery, mortality rate is approximately 90% in the first year of life, resulting from myocardial ischaemia with secondary left ventricular dysfunction, myocardial infarction, mitral insufficiency or fatal arrhythmias.^{1–3} Prognosis and survival to the adult age depend on the development of extensive collateral circulation from the right coronary artery, which characterises the adult type.^{2,3} Early diagnosis allows timely surgical correction and prevention of potential complications.¹ ECG-gated multislice CT angiography is an important diagnostic tool combined with other imaging techniques as it provides a fast and accurate depiction of the origin and course of the coronary arteries. The high spatial resolution and capability of multiplanar reconstructions are essential to the surgical planning.²

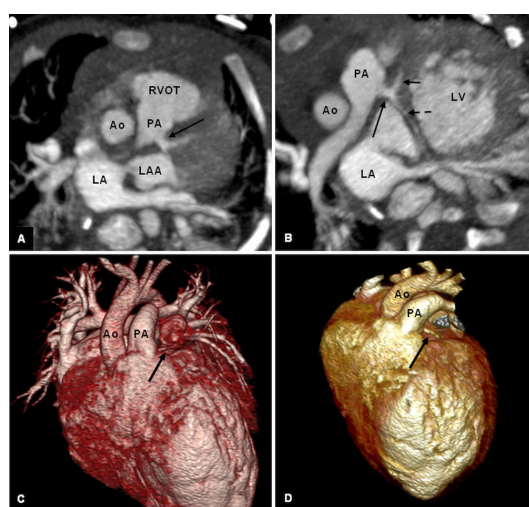


Figure 1 Anomalous left coronary artery from the pulmonary artery (PA) trunk/root junction on multidetector coronary CT angiography. (A) Origin of the left coronary artery (arrow) on the left aspect of PA. (B) The left coronary artery (long arrow) divides into two main arterial branches with course corresponding to the left anterior descending artery (short arrow) and the left circumflex artery (short dashed arrow). (C, D) Volumetric three-dimensional (3D) reconstructions depicting abnormal origin of the left coronary artery (arrow). Ao, aorta; LA, left atrium; LAA, left atrial appendage; LV, left ventricle; RVOT, right ventricular outflow tract.

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