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Clinical vignette

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Bland—White—Garland syndrome: extensive collaterals prevent ischaemia

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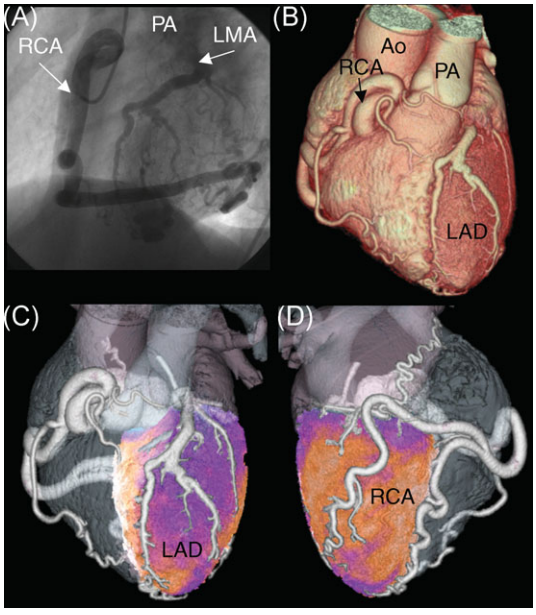
A 56-year-old female patient was referred for coronary angiography because of occasional chest heaviness and a positive stress test. Coronary angiography revealed an anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) also referred to as Bland–White–Garland syndrome. The right coronary artery (RCA) supplied the left coronary system through an abundance of predominant septal collaterals. The left main artery (LMA) drained into the pulmonary artery (PA) (Panel A).

On 64-slice CT angiography (Panel B), the left anterior descending artery (LAD) appeared wrinkled and thin walled, due to the low pressure and retrograde flow into the pulmonary artery. The extensive collaterals resulted in a preserved perfusion at rest and almost normal coronary flow reserve of the anterior wall, as documented by $[^{13}N]NH_3$ positron emission tomography (PET). Nevertheless, fused PET–CT imaging showed a relative hypoperfusion of the anterior wall (purple colour, Panel C) compared with the privileged perfusion of the inferior wall (orange colour, Panel D).

The Bland–White–Garland syndrome is a rare congenital condition. Eighty percent of affected infants die within 4 months. Survival is critically dependent on the development of collateral circulation. In adulthood, this syndrome is seen with angina, congestive heart failure, mitral regurgitation, and sudden death.



Panel A. Coronary angiography showing blood flow from RCA to LMA and drainage into the PA.

Panel B. CT angiography.

Panel C. Fused PET–CT imaging showing a relative hypoperfusion of the anterior wall (purple colour).

Panel D. Fused PET–CT imaging showing a privileged perfusion of the inferior wall (orange colour).