

LETTER

Imaging features of complete congenital atresia of left coronary artery

Keywords Coronary artery anomaly; Congenital heart disease; Coronary artery atresia; Cardiac computed tomography (CT)

Dear editor,

Left coronary artery atresia is a rare coronary abnormality in which there is no left coronary ostium, the proximal left main trunk ends blindly, blood flow arises from the right coronary artery to the left one via small collaterals and retrogradely in at least one of the left-sided arteries. We report the imaging features of a 12-year-old girl with complete congenital atresia of left coronary artery.

A 12-year-old girl had been followed since childhood for progressive aortic stenosis and bicuspid aortic valve. She was referred to us for aortic surgical valvuloplasty. During surgery no left coronary ostium was observed. Cardiac computed tomography (CT) showed absence of viable left main stem, anterior descending and circumflex coronary artery. Several collateral vessels arising from right coronary artery were visualised with no even distal left coronary branch identified. Fibrous tissue was evidenced at CT in the usual anatomic plane of left coronary artery (Fig. 1). Cardiac magnetic resonance imaging (MRI) revealed regional dysfunction, stress induced ischemia and subendocardial fibrosis in anterior and antero-lateral segments of left ventricle. Medical treatment was proposed because bypass revascularisation was not possible due to complete absence of distal left vessels.

To our knowledge this is the first reported case of complete atresia of left coronary artery with no left sided epicardial branches. Ostial atresia is seen particularly in association with cardiac malformations such as pulmonary atresia with intact septum, in which coronary perfusion arises from the right ventricle through sinusoids. Embryological, left ostium atresia may be due to defective incorporation of the coronary bud into the corresponding sinus or defective formation of the proximal part of the vessel lumen. Less than 30 pediatric cases and less than 40 adult cases of left coronary artery atresia have been published but none corresponded to complete atresia of left coronary artery with no left branches present as in our patient [1,2]. Angiographic diagnosis is difficult, particularly in young children with ischemic heart disease, and can only be achieved by demonstrating retrograde perfusion of the atretic vessel from the other coronary artery. Differential diagnosis includes ectopic origin of coronary artery [1,2]. Cardiac CT is the examination of choice in patients with suspected anomalous origin of coronary artery because it provides good visualization of coronary ostia, coronary dominance, angulation from the aortic root, ostial narrowing, length of intramural course, presence of coronary fistulas and relationship with other vascular structure [3,4]. In patients with visualisation of only one coronary ostium, cardiac CT allows excluding ectopic origin as in abnormal origin of left coronary artery from pulmonary artery, abnormal origin of right coronary artery from pulmonary artery or abnormal origin of coronary artery from opposite sinus of Valsalva. In this particular pathological condition of complete atresia of left coronary artery, cardiac CT helps identify the presence of fibrous tissue replacing the normal epicardial vessel. Cardiac CT should be considered as the examination of choice in patients with suspicion of single coronary ostium from aortic sinus.

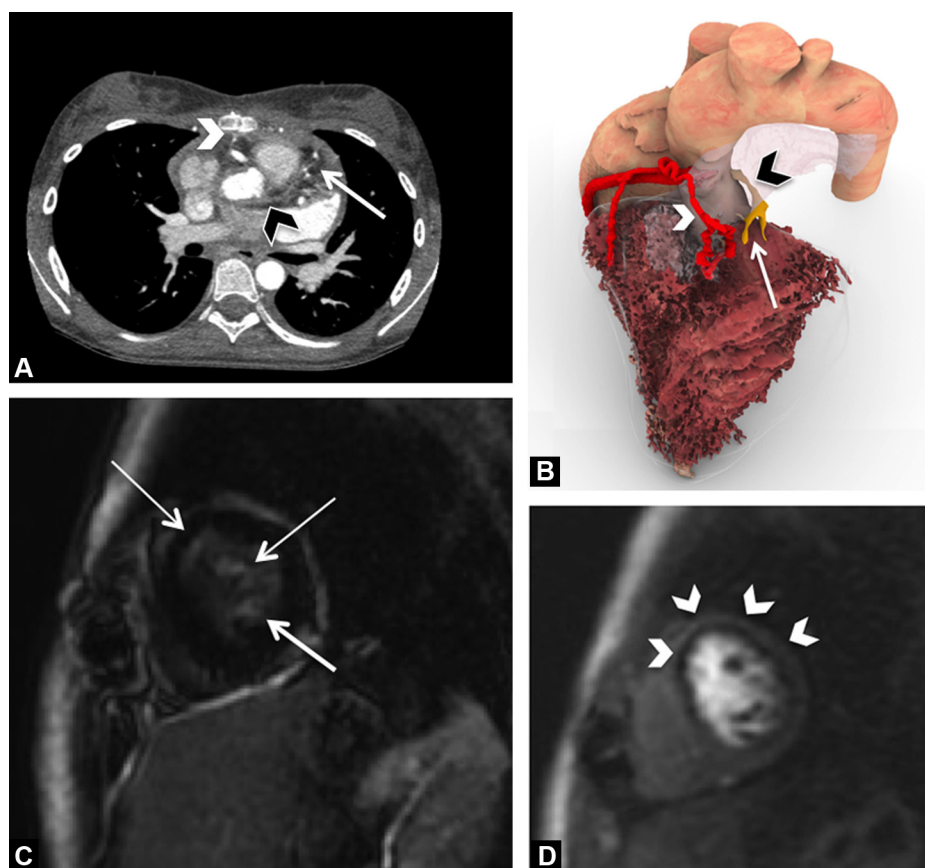


Figure 1. 12-year-old girl with complete congenital atresia of left coronary artery. A. Trans-axial cardiac CT angiography image shows hypoattenuating, unenhanced “atretic” epicardial left coronary artery at the level of left main stem (arrowhead) involving its major branches, Left anterior descending artery (arrow) and a small left circumflex artery. B. Three-dimensional surface rendering reconstruction (Video Supplement) represents the atretic left common artery (LCA) in yellow with its main components: left main stem (black arrowhead) and left descending artery (arrow). Collateral vessels from right coronary artery such as conal branches (white arrowhead) supply LCA territories; of note collateral arteries connect/supply directly to LCA septal ramification rather than epicardial “atretic” branches. C. Post-contrast (Gadoterate meglumine, Dotarem®, Guerbet, 0.2 mmol/kg body weight) cardiac MR image in mid-ventricular short axis reveals subendocardial antero-septal and anterolateral late enhancement involving also papillary muscles (arrows) due to with ischemic myocardial scar. Delayed enhancement images were acquired with an inversion recovery gradient echo pulse MRI sequence (TR/TE = 4.6/1.3 ms; Inversion time [TI] = 240 ms; Flip angle = 20°). D. Cardiac MR perfusion image obtained under pharmacological stress highlights wide inducible myocardial ischemia (arrowheads) involving left common artery territories in mid ventricular short axis view. Perfusion MRI was performed using fast gradient echo single shot time course sequence with the following parameters (22 views/segment for heart rate < 75 bpm or 14 for heart rate > 75 bpm; TR = 3.2–4 ms; Flip angle = 15°). First pass perfusion was evaluated after dipyridamole infusion (0.56 mg/kg over 4 minutes and waiting for heart rate increase > 10%) and repeated at rest.

Authors’ contributions

Raimondi F: Conceptualization, Methodology, Writing- Original draft preparation, Reviewing and Editing.

Scinaro A: Writing- Original draft preparation.

Boddaert N: Supervision.

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Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at <https://doi.org/10.1016/j.diii.2020.02.009>.

Disclosure of interest

The authors declare that they have no competing interest.

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