

Unique family clustering of anomalous left main coronary artery origin from the right sinus of Valsalva: a case for echocardiographic screening and genetic determination

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We report a unique imaging finding of a mother and a daughter with the same type of anomalous left coronary artery origin from the right sinus of Valsalva (ALCAOS) with retroaortic course, and a rarely observed single coronary artery. Anomalous coronary ostium is diagnosed in 1% of angiograms and is not considered hereditary. We present a previously unreported coincidence of anomalous left main coronary artery (LMCA) origin from the proximal right coronary artery (RCA) in a mother and from the right sinus of Valsalva in her daughter.

A 73-year-old woman was admitted to our department with non-ST-segment elevation myocardial infarction and rapid atrial fibrillation. On transthoracic echocardiography (TTE) contractility was normal but anomalous coronary artery was suspected based on the finding of a “bleb sign” and a “crossed aorta sign,” suggesting retroaortic course of the coronary artery (FIGURE 1A and 1B; Supplementary material, *Video S1* and *S2*).¹⁻³ Coronary angiography showed a rare anatomy of a single coronary artery with LMCA originating from the proximal RCA with retroaortic course, without atherosclerotic lesions (FIGURE 1C and 1D). Type 2 infarction was diagnosed; the patient was cardioverted and had 4 years of uneventful follow-up (Supplementary material, *Video S3*).

Four years later her daughter underwent a workup of angina in the setting of type 2 diabetes, rheumatic arthritis, and Graves' disease. She had undergone surgical closure of patent ductus arteriosus at the age of 6. Unexpectedly, the bleb and crossed aorta signs were found on TTE, and

computed tomography angiography confirmed LMCA arising from the right sinus of Valsalva, without atherosclerotic lesions (calcium score of 0). Similarly to her mother, the course of the artery was retroaortic (FIGURE 1D-1H; Supplementary material, *Video S4*). Such clustering of similar ALCAOS including a very rare single ostium variant is unprecedented in the literature.

Anomalous left main coronary artery origin from the opposite (right) sinus of Valsalva is a congenital anomaly reported in 1:600 coronary angiograms, whereas a single coronary artery originating from the RCA is exceedingly rare (1:30 000). Embryology of coronary arteries depends on vasculogenesis and has plausible genetic determinants.⁴ Coronary anatomy in our patients was similar except for the very origin of the LMCA: from the first centimeter of the RCA in the mother, and (separate but adjacent to the RCA) from the right sinus of Valsalva in the daughter. As a chance for a random coincidence of such similar anatomy is no more than 1:18 million, this makes a strong case for potential genetic background of ALCAOS.

We wish to emphasize the possibility of effective TTE screening for retroaortically coursing coronary artery. In previous publications we proposed the names for echocardiographic signs which would help in its identification: a “bleb sign” for short-axis presentation of retroaortic course of the artery and a “crossed aorta sign” for its longitudinal section in modified 5-chamber view; both names were adopted in some later reports.^{1,2,4,5} In a later publication, a term “RAC

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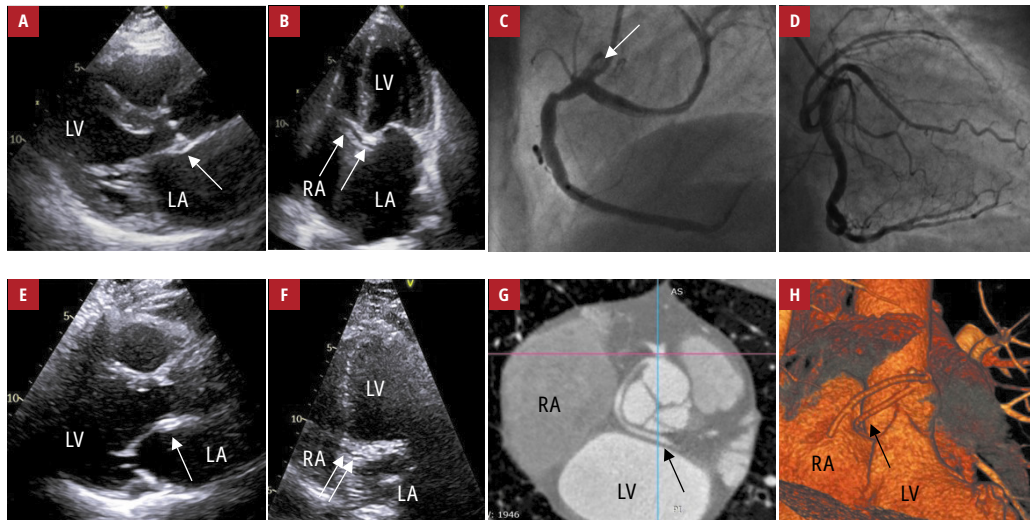


FIGURE 1 **A–D** – mother; transthoracic echocardiography showing a bleb sign (**A**; arrow) and a crossed aorta sign (**B**; arrows); **C** – coronary arteriogram showing the origin of the left main coronary artery (LMCA) from the proximal right coronary artery (RCA); arrow indicates the ostium of the single coronary artery; **D** – selective RCA injection leading to a filling of the entire coronary circulation; **E–H** – daughter; transthoracic echocardiography showing a bleb sign (**E**; arrow) and a crossed aorta sign (**F**; arrows), **G, H** – coronary computed tomography angiography showing the origin of the LMCA 1 cm posterior from the RCA ostium; on both panels the arrow indicates retroaortic course of the left coronary artery
Abbreviations: LA, left atrium; LV, left ventricle; RA, right atrium

sign” (retroaortic anomalous coronary) was also accepted for a similar long-axis presentation of retroaortically coursing coronary artery.⁴ Usually (but not always), the bleb is formed by the circumflex artery originating from the right sinus of Valsalva or the proximal RCA. Confirmation of these signs on TTE encourages to further identify the specific anatomy with computed tomography or coronary angiography, especially if the clinical presentation suggests coronary artery disease, which is not rare in anomalous circumflex artery.²

In Supplementary materials we discussed some additional issues and studies related to the diagnosis and treatment of anomalous left coronary artery.

SUPPLEMENTARY MATERIAL

Supplementary material is available at www.mp.pl/kardiologiapolska.

ARTICLE INFORMATION

CONFLICT OF INTEREST None declared.

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