

Supplementary material

Kasprzak JD, Peruga JZ, Lipiec P, et al. Unique family clustering of anomalous left main coronary artery origin from the right sinus of Valsalva: a case for echocardiographic screening and genetic determination. Kardiol Pol. 2021; 79: 344-345. doi:10.33963/KP.15785

Please note that the journal is not responsible for the scientific accuracy or functionality of any supplementary material submitted by the authors. Any queries (except missing content) should be directed to the corresponding author of the article.

We would like to highlight the utility of multimodality imaging in the detailed diagnostic work-up of anomalous coronary arteries. Beyond the coexisting coronary artery disease, sometimes causing the diagnostic difficulties amid first coronary angiography performed during acute clinical presentation [6], retroaortically coursing coronary artery may coexists with significant valve disease potentially hampering surgical intervention [7,8].

In transthoracic echocardiography the detection of „bleb sign”, contrary to more convenient for this transesophageal echocardiography, requires good resolution and left parasternal window of adequate quality which may be lowered by calcification of mitral annulus or both mitral and aortic valves [7]. On the other hand the visualisation of long axis of retroaortically coursing coronary seems to be easier also in TTE, presenting not only at the level of aorta in five-chamber view, as „crossed aorta sign”, but also in four chamber at right atrium level, where should be differentiated with coronary sinus [7,9]. This TTE presentation of longitudinal section of anomalous artery (both at the level of aorta and right atrium) was described by Witt CM et al. in 2017 as „RAC- retroaortic anomalous artery” sign and tested for sensitivity and specificity in retrospective analysis of 49 cases and 49 controls having done also CT angiography, which rendered 63.3% sensitivity and 93.9% specificity of those sign in the detection of retroaortic coronary artery which may be not only circumflex artery (the most often coronary anomaly) but also left main coronary artery as in our familial case

described in current number of *Kardiologia Polska* [9]. In the literature, there are cases supporting the surgical treatment in the settings of retroaortically going left main coronary artery when causing angina symptoms (e.g case of a 19-year-old man, in whom anomalous coronary anatomy was not suspected amid TTE and neither correctly diagnosed during coronary angiography when intratruncal course were surmised, but the interoperative diagnosis and reimplantation of anomalous artery origin was successfully performed, reference number [10], as well as some reports of myocardial infarctions and sudden cardiac deaths in patients with anomalous origin of left and circumflex coronary artery [10-13].

It is therefore worth emphasising that computed tomography images (CT) offering both 3D and multiplanar reconstructions with high spatial resolution are often the best choice for full delineation of origin, course and location of the anomalous heart vessels as well as their relations to the neighbouring structures, which may be crucial for further monitoring and treatment [14,15].

Additional References:

6. Pawlik A, Januszek R, Dudek D, Bartuś S. A 48-year-old patient with non-ST-elevation myocardial infarction caused by a narrowing in the anomalous circumflex artery. *Pol Arch Intern Med.* 2019; 129: 424-425.
7. Wierzbowska-Drabik K, Peruga JZ, Lipiec P, et al. Transthoracic and Three-Dimensional Transesophageal Echocardiographic Presentation of Anomalous Circumflex Origin from Right Coronary Artery in Patient with Severe Mitral Stenosis. *Echocardiography.* 2015; 32: 1599-1600.
8. Vaishnava P, Pyo R, Filsoufi E, Sharma S. Compression of the anomalous left circumflex artery after aortic and mitral valve replacement. *Ann Thorac Surg* 2011; 92: 1887-1889.
9. Witt CM, Elvert LA, Konik EA et al. The RAC sign: retroaortic anomalous coronary artery visualisation by transthoracic echocardiography. *JACC Cardiovasc Imaging.* 2018; 11: 648-649.
10. Kejriwal NK, Tan J, Gordon SP, Newman MA. Retroaortic course of the anomalous left main coronary artery: is it a benign anomaly ? A case report and review of literature. *Heart Lung Circ* 2004; 13: 97-100.
11. Murphy DA, Roy DL, Sohal M et al. Anomalous origin of the left main coronary artery from anterior sinus of Valsalva with myocardial infarction. *J Thorac Cardiovasc Surg.* 1978; 75: 282-285.
12. Mustafa I, Gula G, Radley-Smith R, Durrer S, Yacoub M. Anomalous origin of the left coronary artery from the anterior aortic sinus: a potential cause of sudden death. Anatomic characterization and surgical treatment. *J Thorac Cardiovasc Surg.* 1981; 82: 297-300.

13. Paterson FK. Sudden death in a young adult with anomalous origin of the left circumflex artery. *S Med. J.* 1982; 75: 748-749.
14. Kwiatkowska J, Herrador Rey A, Meyer-Szary J, et al. Long-term outcome after surgical repair of anomalous origin of the left coronary artery from the pulmonary artery: 24 years of experience. *Kardiol Pol.* 2019; 77: 716-718.
15. Yoshihara S, Yaegashi T, Matsunaga M et al. Anomalous coronary sinus ostium on cardiac computed tomography. Letter to Editor. *Kardiol Pol.* 2020; 78: 947-948.

Supplementary Videos:

Video S1. The cross-section of the retroaortically coursing coronary artery is visible as a „bleb sign” in a standard long axis parasternal view, at the mitroaortical junction.

Video S2. In a slightly modified apical five-chamber view the part of longsection of retroaortically placed coronary artery is displayed forming „crossed aorta sign”.

Video S3. Coronary angiography in mother showing a single coronary artery with left main coronary artery originating from the proximal right coronary artery.

Video S4. 3D reconstruction of computed tomography displaying left main coronary artery arising from the right sinus of Valsalva.