Anomalous origin of the circumflex artery from the right Valsalva sinus on transthoracic echocardiography

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A 62-year-old woman with a history of hypercholesterolaemia, asthma, and radiofrequency ablation of right ventricular outflow tract arrhythmia presented to an outpatient cardiology clinic for a scheduled follow-up visit. An additional tunnel-like echo, suggestive of anomalous origin of the coronary artery, was found on echocardiography (Fig. 1A, Suppl. Video 1, 2 — see journal website). The patient was asymptomatic for arrhythmia or ischaemia, and no abnormalities were recorded on an electrocardiogram; therefore, no further evaluation with computed tomography angiography was conducted. A similar finding was observed in our echo-lab during cardiac ultrasonography in a 70-year-old man with a history of ischaemic stroke, hypertension, and hypercholesterolaemia (Fig. 1B, Suppl. Video 3, 4 — see journal website). The patient had symptoms of angina and was referred for coronary angiography, which revealed a multivessel disease. Finally, he was scheduled for coronary artery bypass graft surgery. The patient’s circumflex artery (Cx) originated from the proximal segment of the right coronary artery (RCA) (Fig. 1C, D, Suppl. Video 5, 6 — see journal website), which rendered a similar tunnel-like structure visible on echocardiography. Congenital anomalies of coronary arteries are a heterogeneous group of disorders with various manifestations. They are present in about 1% of patients undergoing coronary angiography [1], but the actual incidence may be higher. According to AHA/ACC, it occurs in subjects with an anomalous origin of a coronary artery from the opposite Valsalva sinus, typical symptoms either do not occur at all or may include chest pain, dyspnoea, palpitations, syncope, cardiomyopathy, arrhythmias, and myocardial infarction or sudden cardiac death (SCD) — the latter occurring predominantly among young patients after extreme exertion [1]. Available data suggest an increased risk of SCD among patients presenting with the left coronary artery arising from the right Valsalva sinus and coursing between the pulmonary artery and aorta. A screening protocol to prevent SCD in athletes and other young individuals subjected to extreme exertion presenting with atypical chest pain, syncope, or dyspnoea should comprise both echocardiography (including Holter monitoring) and echocardiographic evaluation with identification of the origin of the coronary arteries. In elderly patients, recognition of the anomalous origin of the coronary artery on echocardiography is also useful. This was shown in a review of more than 10,000 angiographic records, where the origin of Cx from the opposite side of the aorta was related to coronary atherosclerosis in the proximal portion of the anomalous vessel. This means that the posterior course of the anomalous Cx (Fig. 1E, F) may predispose this vessel to atherosclerosis [1]. When coronary artery angiography is planned in patients with this condition, it may lead to a false diagnosis of Cx absence, if not visualised as a branch originating from the left main coronary artery. Moreover, it can cause potential technical challenges during percutaneous coronary interventions (PCIs) (problems with optimal catheter selection, difficulties with wire manoeuvring, balloon or stent delivery), which may lead to an increased risk of PCI failure or periprocedural complications. In summary, the anatomical variant of Cx origin from the right Valsalva sinus can be at least suspected on the basis of transthoracic echocardiography. This information is useful in both symptomatic young individuals and in older patients. Undoubtedly, having seen this unusual artery anatomy and course at least once is helpful to avoid missing this anomaly on echocardiogram. Hopefully, the presented case helps clinicians in diagnosing the not so rare and not so trivial anomalous origin of the circumflex branch from the right Valsalva sinus.

References

Figure 1. The additional tunnel-like echo, suggesting anomalous origin of the coronary artery; first (A) and second (B) patient. Right coronary artery (RCA, red arrow) and circumflex artery (Cx, yellow arrow) in left anterior-oblique 30° view (C) and postero-anterior 0° view (D). Diagrams showing normal (E) and anomalous (F) origin of Cx; L — left; LAD — left anterior descending; R — right; N — non-coronary sinus; PA — pulmonary artery

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