ДРУЖЕСТВО НА КАРДИОЛОЗИТЕ В БЪЛГАРИЯ
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# ЕХОКАРДИОГРАФИЯ НА КОРОНАРНИ АРТЕРИИ ПРИ ВЪЗРАСТНИ И РОЛЯТА Ѝ В МУЛТИМОДАЛНАТА ОБРАЗНА ДИАГНОСТИКА: ПРИМЕРИ ОТ РАЗЛИЧНИ КЛИНИЧНИ СЦЕНАРИИ И КЛИНИЧЕН СЛУЧАЙ

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# CORONARY ARTERY ECHOCARDIOGRAM IN THE ADULT PATIENT AND ITS ROLE IN MULTIMODALITY IMAGING: EXAMPLES OF DIFFERENT CLINICAL SETTINGS AND A CASE REPORT

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**Резюме.** Вродените аномалии и придобити патологии на коронарните артерии все по-често се идентифицират посредством различни образни методики. Анормалният аортен произход на коронарна артерия от противоположния синус се оценява на 0,7%. Ехокардиографията може да има инкрементална стойност при тези сложни пациенти, които често изискват мултимодален диагностичен подход. Представяме случай на пациентка с единствена коронарна артерия, идентифицирана чрез ангиография, с курс на ствола на лявата коронарна артерия между аортата и изходния тракт на дясната камера. Също така накратко изброяваме редица различни клинични сценарии при пациенти със структурно нормални сърца, с вродени сърдечни заболявания и пациенти след сърдечна операция или интервенции. Те са илюстрирани с различни ехокардиографски, ангиографски и томографски изображения.

**Ключови думи:** ехокардиография на коронарни артерии, коронарни аномалии, мултимодална образна диагностика

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**Abstract.** Congenital coronary artery anomalies and acquired coronary artery pathologies are increasingly identified by different imaging modalities. Abnormal aortic origin of a coronary artery from the opposite sinus is estimated at 0.7%. Echocardiography may have an incremental value in these complex patients often requiring multimodality diagnostic approach. We present the case of a female patient with a single coronary artery identified by angiography with left main course between the aorta and right ventricular outflow tract. We also briefly list a number of different clinical scenarios in patients with structurally normal hearts, with congenital heart disease and patients after cardiac surgery or interventions. These are exemplified by different echocardiographic, angiographic and tomographic images.

**Key words:** coronary artery echocardiogram, coronary artery anomalies, multimodality imaging

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## Introduction

Older observational data suggest that LAD originating from the right sinus of Valsalva is a rare anomaly with a prevalence of 0.03% and left main artery originates from the right sinus in 0.017% [1]. More recent analyses estimates that abnormal coronary origin from the opposite sinus occurs at 0.7% [95% CI: 0.48-0.95%] with right coronary artery (RCA) origin from left sinus being six times more prevalent than left coronary artery (LCA) origin from the right sinus [2]. As in some cases coronary anomalies could be associated with el-

evated risk of sudden cardiac death (SCD), better recognition and detailed imaging evaluation are becoming a necessity. Coronary artery imaging by echocardiography is most often achievable in short axis view above the level of aortic cusp coaptation. Rotating the probe clockwise may help in better delineation of LCA and counterclockwise rotation facilitates imaging of RCA. In experienced hands (and with appropriate patient habitus) coronary artery course could be identified caudal to the aortic root in apical views (the so called "Retro-Aortic Coronary or RAC" sign) and more rarely in the anterior or posterior interventricular sulcus.

## Case description

A 52-year-old normotensive female patient with a history of seropositive rheumatoid arthritis was evaluated in 2017 in another institution because of atypical chest pain and undue fatigue. She had a normal transthoracic echocardiogram finding, but invasive coronary angiogram revealed a single coronary artery, arising from the right sinus with a presumed "pre-pulmonary course" (Fig. 1). She was admitted to our institution for multi-detector computer tomographic coronary angiogram. Her laboratory results were unremarkable and her ECG and Holter ECG were normal.

Echocardiography revealed normal left ventricle mass, normal left and right ventricle dimensions and systolic function without any segmental wall motion abnormalities. The common coronary artery was identified arising from the right sinus of Valsalva dividing into a large-caliber RCA and a small-caliber LCA (Fig. 2). The color flow in LCA could be identified by a parasternal long axis view (Fig. 2, http:/10.3897/bgcardio.30.e141038.Videofiles). A course of LCA between the aorta and the right ventricular outflow tract (and towards the interventricular septum) was defined by echocardiography.

A MDCT was performed and confirmed that LCA has a subpulmonary or intraseptal course (Fig. 3) as opposed to intraarterial course which is associated with elevated risk of SCD. Both computed tomography and the invasive coronary angiogram demonstrated the so-called "hammock sign" which is a characteristic feature

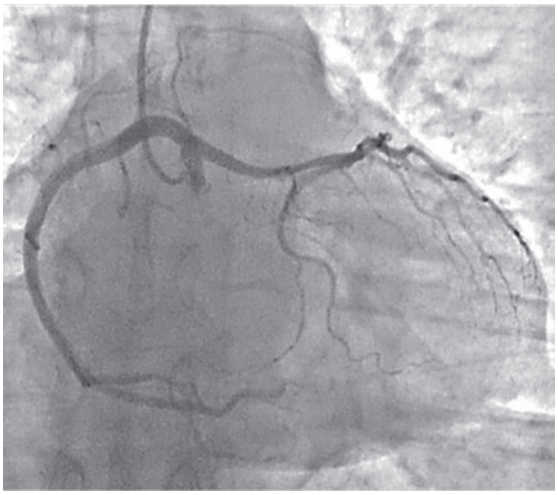

**Fig. 1.** Coronary angiogram of a single coronary artery arising from the right sinus

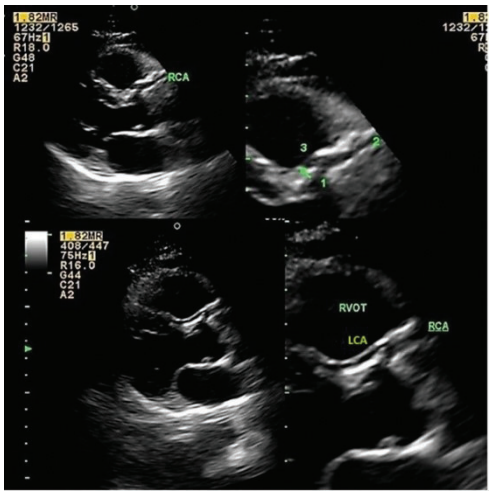


**Fig. 2.** Long axis view demonstrating LCA course between the aortic root and RVOT

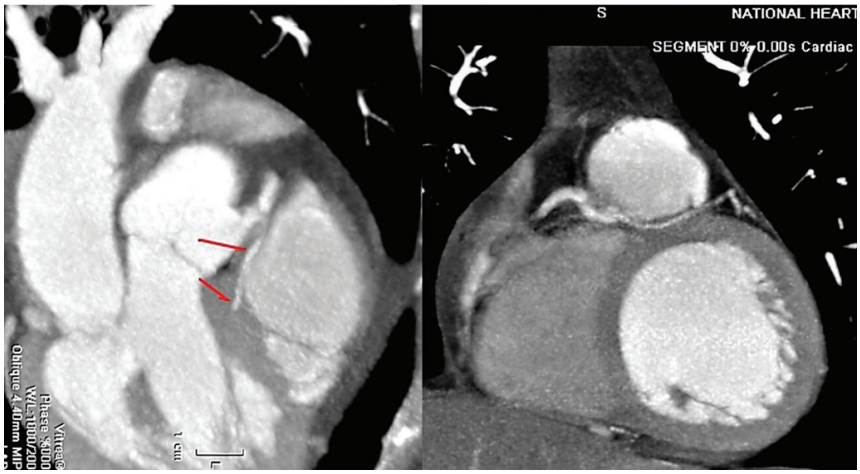


**Fig. 3.** Computed tomography demonstrating the so-called "hammock" sign with subpulmonary or "intraseptal" course of LCA

of intraseptal artery position and is expected to have a benign clinical course [3].

A $^{99}$mTc MIBI – TF SPECT/CT-stress test was performed, using bicycle with Bruce protocol. The test was stopped after only 50Watts due to muscle fatigue and dyspnea. No ischaemic ECG changes were recorded, there was a slight and nonsignificant anteroseptal hypoperfusion with extent of 4-5%. The segmental kinetics, left ventricle volumes and ejection fraction were normal at baseline and with exercise (LVEF 74%) and Ca score was 0 (zero) (Fig 4).

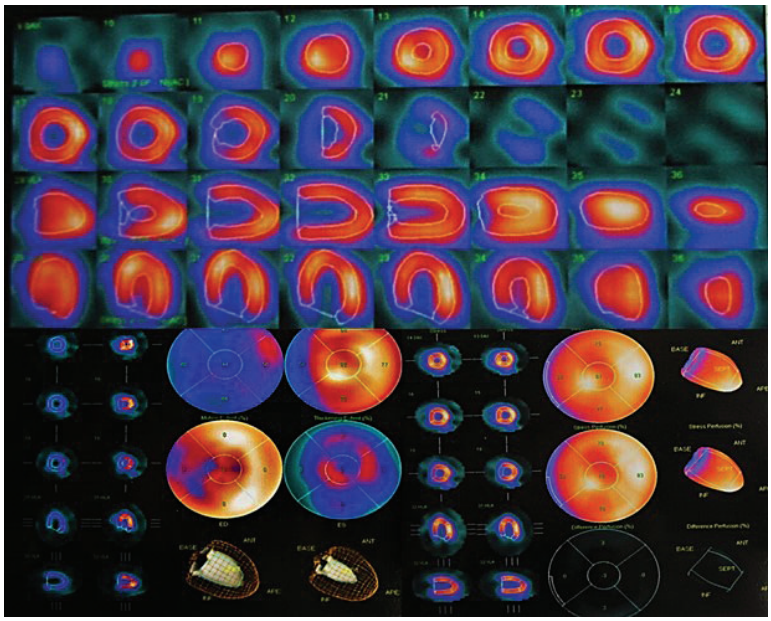

**Fig. 4.** Myocardial perfusion scintigraphy with 99mTc-MIBI

The patient was discharged with low dose beta blocker and was regularly followed up with clinical visits and echocardiography. With time she developed arterial hypertension, and a calcium channel blocker was added to the therapy. By the time of this manuscript submission her clinical course has been uneventful for 7 years.

## The role of coronary echocardiogram in different clinical settings:

Except in the presented clinical case, we have encountered several different clinical scenarios in which coronary artery echocardiogram was more than an imaging curiosity and has often required making important decisions.

We were able to identify by transthoracic echo different types of coronary origin anomalies, including RCA from the left sinus of Valsalva (Fig. 5, http:/10.3897/bgcardio.30.e141038.Videofile5) (which shares common diagnostic and treatment challenges as the presented case of LCA from the right sinus), retroaortic coronary arteries which are usually associated with benign clinical course and even dual left anterior descending (LAD) coronary arteries (Fig. 6) or dual LCx coronary arteries with the abnormal circumflex artery originating from the right sinus and coursing behind the aortic root (Fig. 7, http:/10.3897/bgcardio.30.e141038.Videofile2).

Another clinical scenario that we have faced is the necessity of complete non – invasive imaging evaluation in patients with grown-up congenital heart disease even if cardiac catheter-

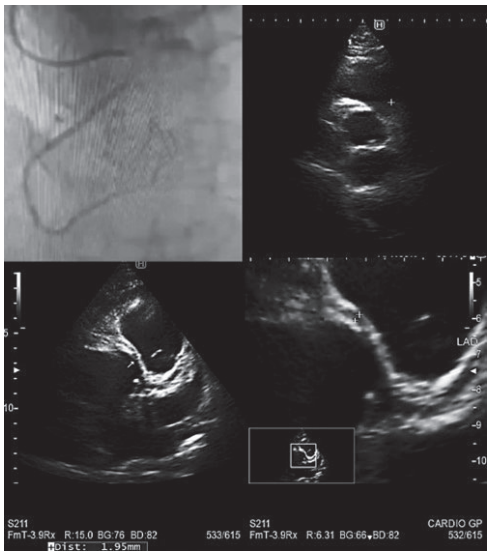

**Fig. 5.** Coronary angiogram revealed abnormal RCA origin and echocardiography identified it to be from the left sinus with initial intramural course. Computed tomography confirmed this diagnosis and the patient was referred to as stress scintigraphy

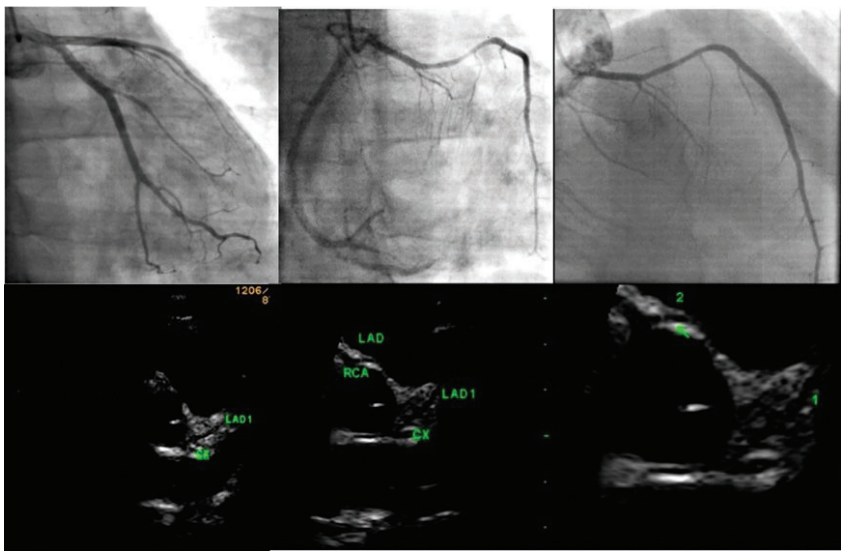


**Fig. 6.** 37-year-old male with pericarditis, fever and elevated CRP and troponin. Invasive angiogram identified dual LAD with the abnormal artery arising from the right sinus exhibiting coronary spasm. **Top left** – small-caliber LAD from left sinus, **Top right** – resolution of coronary spasm with nitrate. **Bottom**: short axis echocardiographic images. LAD 2 could be mistaken for large conus branch, but its course was visualized also in PLAX view

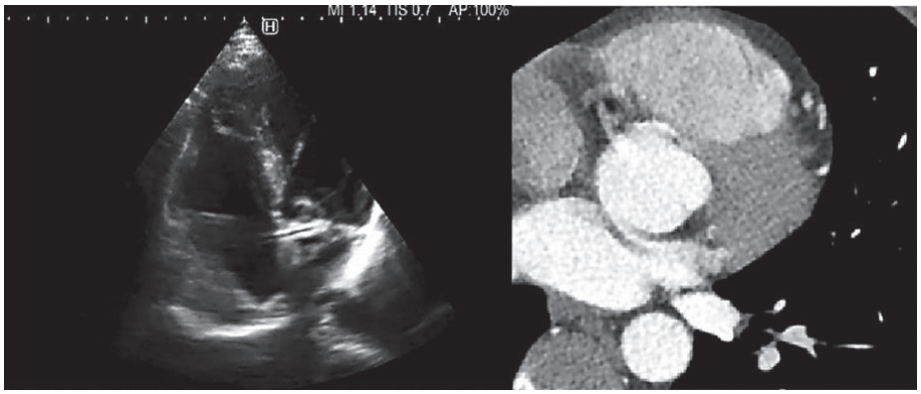

**Fig. 7.** 47-year-old male with borderline high blood pressure and dyslipidemia, presenting with atypical chest pain. Left – "RAC" sign caudal to the aortic root in apical view. Computed tomography revealed dual LCx artery with abnormal small caliber branch with a course behind the aorta. No coronary stenoses were identified

ization is planned. Computed tomography can provide irreplaceable clinical information but echocardiography should be the first imaging modality – thus the most appropriate imaging protocol for MDCT (opacification of right or left chambers, coronary arteries or extracardiac collateral anastomoses) or CMR (quantification of chamber volumes and function, myocardial LGE, shunts, regurgitant fractions) might be selected. Here we present a complex case of a 42-year-old male with normal coronary arteries identified before cardiac catheterization by transthoracic echocardiography (Fig. 8) and a 42-year-old female patient with double outlet right ventricle, ventricular septal defect and transposition of the great arteries (Fig. 9, http:/10.3897/bgcardio.30.e141038.Videofile4).

Coronary artery echocardiography may have incremental value in the diagnostic evaluation of patients who have undergone cardiac surgery. Here we present an example of coronary stents implanted in RCA and LCA in a patient with type I dissection, ascending aortic vascular graft and multiple descending aorta stent-grafts, all of them implanted in other institutions (Fig. 10, http:/10.3897/bgcardio.30.e141038.Videofile3). The other example is of a patient with ascending aorta graft, enwrapped in the native aorta. Due to dehiscence in front of the reimplanted RCA there was pulsatile high-pressure flow between the graft and the native aorta that was identified by echocardiography (Fig. 11).

In a 59-year-old male patient with ischaemic heart disease and coronary artery bypass surgery

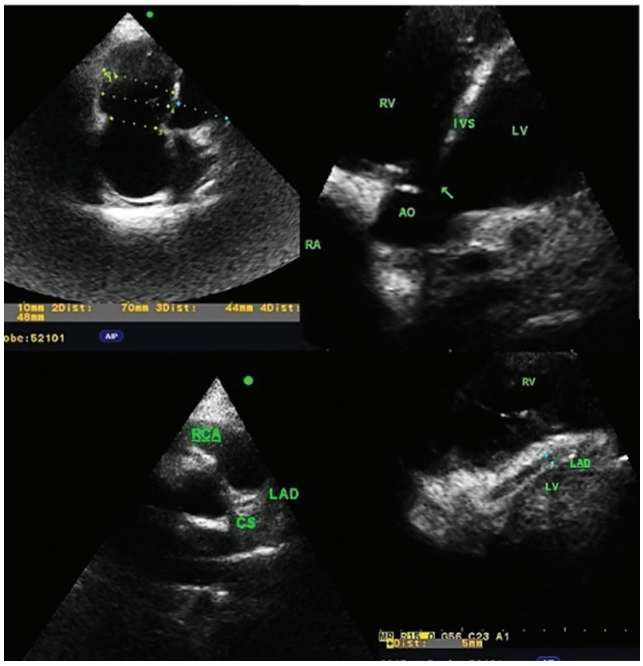


**Fig. 8.** A 42-year-old male with ventricular septal defect, subvalvular pulmonary stenosis and right inflow enlargement. Upper left: severely enlarged right ventricle, upper right – VSD, lower left – short axis view of biostial coronary anatomy, lower right – LAD coursing in the anterior interventricular sulcus

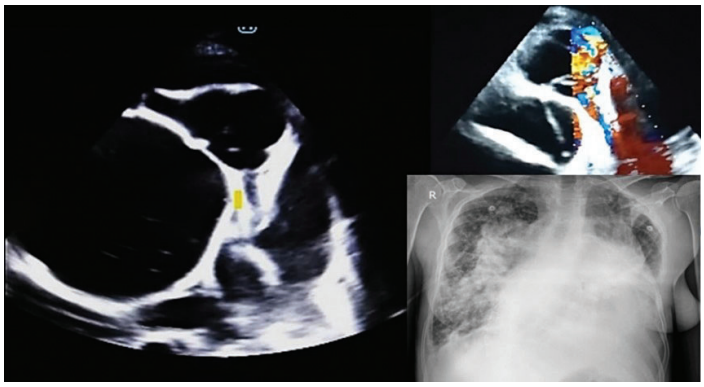

**Fig. 9.** A 42-year-old female with double outlet right ventricle, VSD and transposition of the great arteries. The anterior vessel is identified as aorta with coronary arteries exiting the posterior facing sinuses. The posterior vessel is the giant pulmonary artery

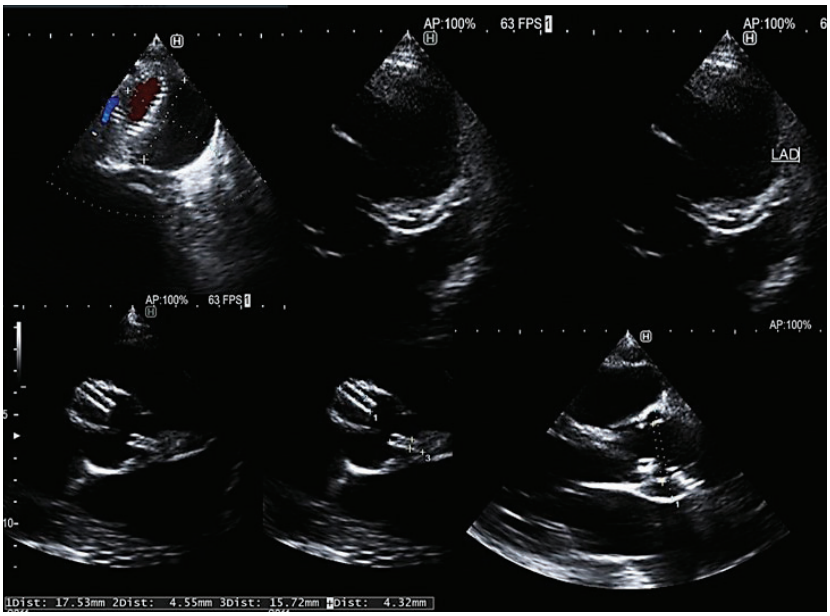


**Fig. 10.** A female patient with ascending aorta graft and multiple descending aorta stent-grafts. Coronary arteries were reimplanted, and the graft and later ostial stents were implanted. Upper left: abdominal aorta short axis view with pulsatile flow within the stent-graft. Middle and upper right – LAD was visualized widely patent. At the bottom – stents in LCA in RCA protruding in the aorta, bottom left – stent in PLAX at the level of the aortic valve


**Fig. 11.** This patient had ascending aortic graft wrapped in the native aorta with coronary artery reimplantation. Due to dehiscence in front of RCA there was high-pressure pulsatile flow between the graft and the native aorta

in 2002, an invasive coronary angiogram identified large abnormal formation without distal opacification of the venous graft to RCA communication (Fig. 12). An echocardiography from atypical right parasternal (3 to 4th intercostal space) revealed a large aneurysm with pulsatile flow and mural thrombosis (Fig 13). In a report from a single institution after review of more 5500 grafts, only 0.07% graft aneurysms were found [17]. The clinical course, diagnosis and treatment of graft aneurysms are beyond the scope of this article.

Another important scenario to have in mind is identifying the relations of coronary artery to cardiac and extra-cardiac tumors, masses etc. (postoperative abscesses, paravalvular abscesses, mediastinal tumors, myocardial or epicardial cardiac tumors). It requires multimodality imaging approach, but echocardiography may still reveal easily accessible morphological and hemodynamic findings (for example compression or valve obstruction). Here we present the case of a 32-year-old female patient with large intramyocardial hydatid cyst. LAD and proximal LCx patency were visible by echo-

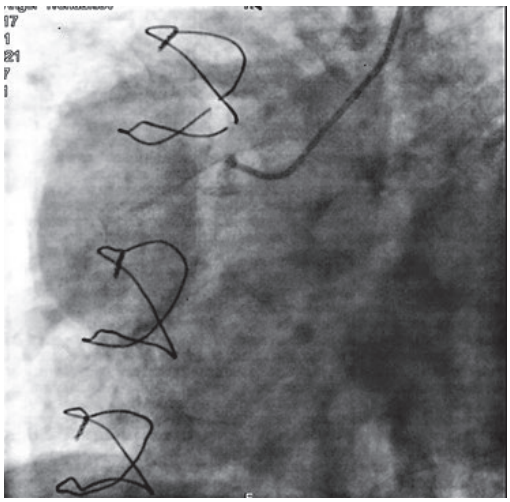

**Fig. 12.** A large venous graft formation in 59 years male after CABG

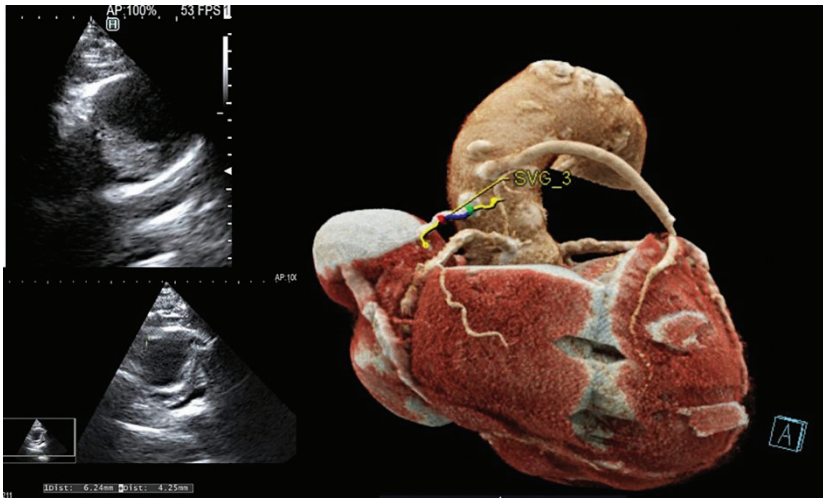


**Fig. 13.** Large graft aneurysm with echographic and CT evidence of partial thrombosis. Volume rendering 3 D heart image (created by S. Dineva)

cardiography. MDCT identified cranial displacement of the LCx and additional intermedial left branch coursing above the formation (Fig. 14).

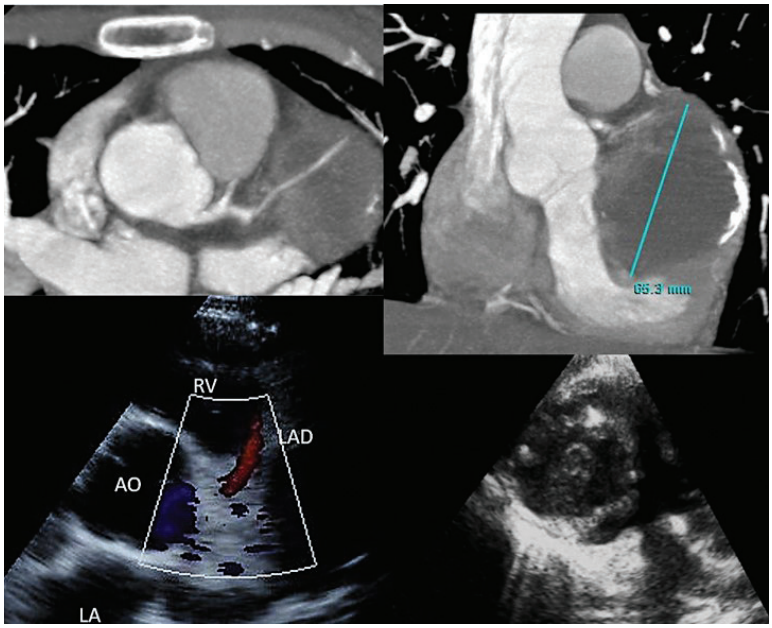


**Fig. 14.** 32-year-old female with a cardiac hydatid cyst in the left ventricle free wall. Top: CT revealed intermedial left branch coursing above the cyst. Bottom left: LAD flow easily seen because of cranial coronary dislocation. Bottom right: atypical position revealing the whole hydatid cyst

## Discussion

In the literature the finding of anomalous coronary artery from opposite sinus (ACAOS) has been associated with SCD mainly with exercise [4]. Stress tests have failed to identify patients who later suffered SCD. According to Angelini et al. 80% of the coronary anomalies found in SCD victims consist of an artery with inter-arterial course between the aorta and the pulmonary artery [5]. A 25-year review of autopsies in 18 to 35-year-old US military recruits revealed that from 64 cardiogenic causes of death, 33% (21 cases) were due to ACAOS [6]. In another analysis of 162 consecutive young (below 40 years old) victims of SCD, coronary artery anomalies were estimated to result in sudden death risk of 0.61% (1:161) [7]. It is believed that intense physical exercise results in coronary artery compression and acute ischemia [8]. Other clinical manifestations of ACAOS may include angina or atypical chest pain, shortness of breath, dizziness, palpitations or syncope [9]. Using IVUS (intravascular ultrasound) as well as necropsies have identified that in ACAOS there is often intramural course of the proximal abnormal artery segment which is believed to play a role in the pathogenesis of SCD. It should be noted that some authors claim that sudden death associated with such a coronary anomaly occurs only in the young [5].

Echocardiographic diagnosis of coronary artery anomalies has been of interest for more than 25 years. From 1997 to 2002 Fromelt et al. analyzed 2388 echocardiograms that were able to identify ACAOS in 0.17% of patients as opposed to 1.07% identified by coronary angiography [10]. One patient who was not identified by echocardiography later suffered sudden cardiac death [10]. It should be noted that all patients in this review were children or adolescents in whom coronary artery imaging was more easily achieved compared to adults [11]. According to the American Society of Echocardiography recommendations for multimodality assessment of congenital coronary anomalies, “TTE (transthoracic echocardiogram) should be the initial screening tool for suspected AAOCA, particularly in forms that are associated with myocardial ischemia (those with an interarterial course)” [11]. When an abnormal coronary artery is identified computed tomography and/or cardiac magnetic resonance are recommended for detailed delineation of coronary artery course and origin [11]. More recent echocardiographic data showed that using a standardized protocol increased coronary artery anomalies detection from 0,02% to 0.22% [12]. Yet 6-10% of echocardiograms were excluded in other studies due to uninterpretable image quality [13]. Even after exclusion of these patients Pellicia et al. were unable to identify RCA ostium in 20% of young athletes, according to a publication from 1993 [13].

Understanding the best treatment strategy in coronary artery origin anomalies is still evolving and under debate. According to the ESC Guidelines for congenital heart disease from 2020 , surgery is the recommended treatment for anomalous coronary artery in patients with typical angina and stress induced ischemia attributable to the anomaly (Class I, Level of evidence C) [14]. In asymptomatic patients with inducible ischemia or without proven ischemia but with high-risk interarterial course of LCA, surgical treatment should be considered (Class IIa, Level of evidence C) [14]. Although surgical treatment is the only recognized choice according to the 2015 American College of Cardiology/American Heart Association scientific statement update for Athletes with cardiovascular disease [15], a growing number of studies shows that percutaneous intervention might be a valid option. In a study of predominantly adult patients (mean age 48, n = 42) with anomalous right coronary artery, coronary intervention resulted in a 13% incidence of in-stent restenosis with serial IVUS assessment [16]. In this study 29% of patients had symptom recurrence during a mean follow-up of 5 years [16].

## Conclusion

Congenital or acquired coronary artery pathology often necessitates multimodality imaging for detailed diagnosis and careful treatment plan. The physician should always be aware of the myriad technical and anatomical limitations of coronary echocardiographic imaging. Despite that in experienced hands echocardiography may provide not only coronary ostia identification but sometimes may have incremental diagnostic value. Echocardiography is still the most unexpensive, accessible and fast to perform imaging modality that can provide structural and also hemodynamic data.

*No conflict of interest was declared*

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