Cardiology in the Young

cambridge.org/cty

A rare coronary artery anomaly in a 14-year-old boy with sudden cardiac arrest

Sarah Ruffing[1], Hashim Abdul-Khaliq[2] and Martin Poryo[3]

[1]Department of Pediatric Cardiology, Universität des Saarlandes - Campus Homburg, Homburg, Germany; [2]Department of Pediatric Cardiology, Saarland University Hospital and Saarland University Faculty of Medicine, Homburg, Saarland, Germany and [3]Department of Pediatric Cardiology, Saarland University Medical Center, Homburg/Saar, Germany

Brief Report

Cite this article: Ruffing S, Abdul-Khaliq H, and Poryo M (2023) A rare coronary artery anomaly in a 14-year-old boy with sudden cardiac arrest. *Cardiology in the Young* **33**: 829–831. doi: 10.1017/S1047951122002967

Received: 2 May 2022
Revised: 9 August 2022
Accepted: 25 August 2022
First published online: 22 September 2022

Keywords:
Coronary artery anomaly; cardiac arrest; resuscitation; implantable cardioverter defibrillator

Author for correspondence:
Sarah Ruffing, Department of Pediatric Cardiology, Universität des Saarlandes - Campus Homburg, Homburg, Germany.
E-mail: sarah.ruffing@uks.eu

Abstract

Congenital coronary artery anomalies represent a rare cause for cardiac arrest in children and adults; however, most of these anomalies are asymptomatic and incidental findings. We report on a 14-year-old boy who was admitted to our hospital after cardiopulmonary resuscitation at home. Diagnostic workup including histopathology revealed parvovirus B19 in endomyocardial biopsy. Moreover, cardiac catheterisation as well as CT angiography identified an anomalous origin of the right coronary artery with an interarterial course. Since this anomalous coronary artery might have caused impaired myocardial perfusion causing cardiac arrest, surgical correction and implantation of a cardioverter defibrillator were performed. The further post-operative clinical course (7 months) has been uneventful.

The prevalence of coronary artery anomalies in the general population is estimated at 1.2%. In most cases, these anomalies are asymptomatic and incidental findings. In about 20% of coronary artery anomalies, serious life-threatening symptoms occur, that is chest pain, myocardial infarction, cardiac arrhythmias or sudden cardiac death.

Coronary artery anomalies are classified into anomalies of origin, course, and termination. The anomalous origin of a coronary artery from the opposite sinus of Valsalva is of particular importance because of its strong association with sudden cardiac death. Especially in young athletes suffering from sudden cardiac death coronary artery anomalies play an important role.[1]

Case report

We report on a 14-year-old boy who was admitted to our hospital after cardiopulmonary resuscitation at home. He was found unconscious in his bed by his parents, who started cardiopulmonary resuscitation until arrival of emergency medical service. Initial cardiac rhythm was asystole which converted into ventricular fibrillation. Overall, four direct current defibrillations were administered before return of spontaneous circulation after approximately 25 min. At admission, the patient was cardiopulmonary stable and in sinus rhythm with bigeminal pre-mature ventricular contractions. During in-patient hospital stay, he had alternating pre-mature ventricular contractions (bigeminy, trigeminy) as well as non-sustained ventricular tachycardias, which required beta-blocker therapy. Initial echocardiography revealed no pathological findings, but conditions during examination were impaired.

Medical and family history was unremarkable, especially no sudden cardiac death occurred. The patient had not recently been vaccinated against SARS-CoV-2. He regularly consumed a lot of caffeine, but no illicit drugs or alcohol use was reported.

Diagnostic workup excluded differential diagnoses like arrhythmogenic right ventricular dysplasia and Brugada syndrome. Endomyocardial biopsy revealed intermediate chronically active lymphocytic myocarditis with parvovirus B19, which was interpreted as in the process of recovery. MRI was not applicable because of artificial late enhancement after endomyocardial biopsy. Cardiac catheterisation identified an anomalous origin of the right coronary artery. Cardiac CT angiography was performed to determine the exact course of the right coronary artery (Figs 1 and 2). It originated from the anterolateral left aspect of the ascending aorta 10 mm cranial of the sinotubular junction. The further course tended rightwards around the ascending aorta and then between the pulmonary trunk and ascending aorta. The remaining course was typical in the right atrioventricular groove. The size of the right coronary artery was normal (4 mm, z-score + 2.6), without stenoses or compressions.

Since this anomalous coronary artery might have caused intermittent myocardial hypoperfusion causing cardiac arrest, we decided to transfer the right coronary artery to its usual origin by cardiac surgery. After institution of cardiopulmonary bypass, the right coronary artery was carefully dissected and excised. A new opening was created in the aorta at a more physiologic position, and the right coronary artery was implanted. Moreover, because of

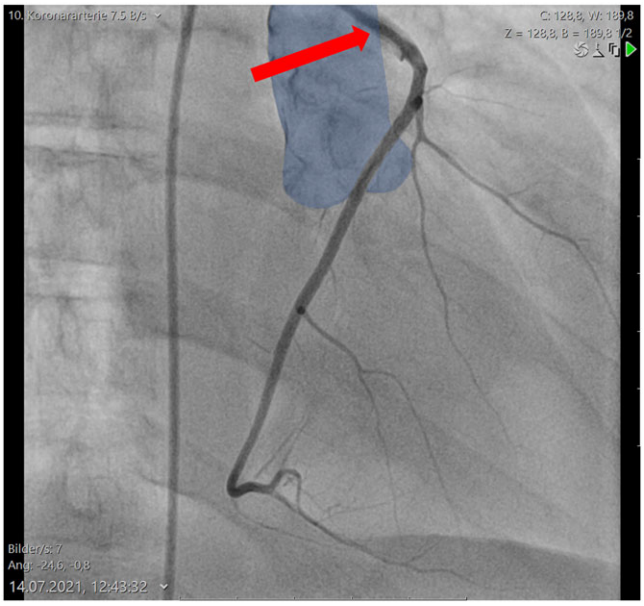

Figure 1. Cardiac catheterisation illustrating the anomalous origin of the right coronary artery (red arrow) 10 mm cranial of the left sinus of Valsalva. Blue marking depicts aortic valve and ascending aorta.

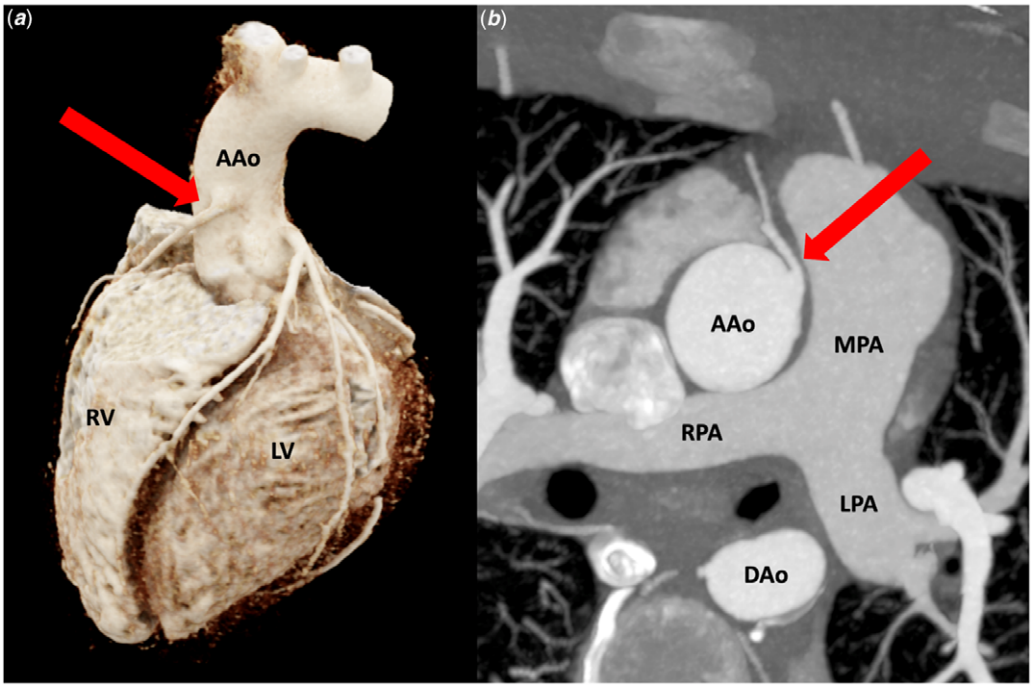

Figure 2. CT angiography illustrating the atypical origin and course of the right coronary artery (red arrows) between ascending aorta and pulmonary artery.

persistent episodes of ventricular tachycardia an implantable cardioverter defibrillator was implanted.

The surgical procedure and the post-operative course were uneventful. During 7-month follow-up, the patient did not report any complaints, the electrocardiography demonstrated a few pre-mature ventricular contractions but was otherwise normal, and the implantable cardioverter defibrillator did not detect any ventricular fibrillation. However, the young patient was diagnosed with arterial hypertension requiring medical treatment.

Discussion

Most coronary artery anomalies are asymptomatic and incidental findings during echocardiography, cardiac CT, MRI, or catheterisation for others reasons, which makes it difficult to determine the real-life prevalence of this congenital malformation. As described previously, the prevalence is thought to be about 1% with a broad distribution of 0.2 - 8.4%. Symptoms like chest pain, myocardial infarction, and cardiac arrhythmias are possible symptoms of coronary artery anomalies but often sudden cardiac death

https://doi.org/10.1017/S1047951122002967 Published online by Cambridge University Press

is the only clinical feature. However, pathophysiologic mechanisms leading to this serious event remain elusive, and there are no specific screening tests for detection of coronary artery anomalies.[2]

In our patient, a right coronary artery originating from the anterolateral left aspect of the ascending aorta 10 mm cranial of the sinotubular junction with an interarterial course was found. Overall, an anomalous coronary artery from the opposite sinus is very rare and found in only about 1% of coronary artery anomalies. They can be differentiated based on its course as interarterial, pre-pulmonic, retroaortic, retrocardiac, and subpulmonic. The interarterial course is the subtype most strongly associated with sudden cardiac arrest/death, especially in anomalous left coronary arteries. It is not for sure whether cardiac arrest in our patient occurred because of the anomalous origin and course of the right coronary artery, myocarditis, or the combination of both. However, results published by Lee et al. support that in this case the high interarterial course of the right coronary artery might have been the leading pathology and myocarditis was only an incidentally finding.[3]

Patients with coronary artery anomalies are not necessarily at need for surgery. The current guidelines state that surgery should be considered in case of anomalous origin of a coronary artery with evidence of myocardial ischaemia as well as in case of anomalous origin of a coronary artery without evidence of myocardial ischaemia but high-risk anatomy.[4] In addition, it is reported that patients with anomalous origin of the right coronary artery from the opposite sinus of Valsalva and high interarterial course are at increased risk of angina pectoris and major adverse cardiac events. Due to cardiac arrest in association with an anomalous origin of the right coronary and high-risk anatomy in our patient, we decided to transfer the right coronary artery to its origin. Furthermore, because of recurrent episodes of ventricular tachycardia we concluded that an implantable cardioverter defibrillator would be the most adequate clinical approach.

Enterovirus and adenovirus are the most common causes of myocarditis in children, but parvovirus B19 is gaining importance as a relevant pathogen in viral myocarditis in this cohort. However, the clinical relevance of parvovirus B19 in this case is uncertain since parvovirus B19 can also be found in tissues of healthy adults without cardiomyopathy or myocarditis in up to 85%.[5]

We conclude that in patients with sudden cardiac arrest during rest and sleep, differential diagnoses should also include coronary artery anomalies. Although myocarditis might cause severe myocardial damage and sudden cardiac death, it is important to realise that not all histopathological findings in viral myocarditis are of clinical relevance. Furthermore, this case report illustrates the importance of rare causes of sudden cardiac collapse in the paediatric population.

Acknowledgement. Special thanks to Jochen Pfeifer, Peter Fries, Felix Mahfoud, and Sascha Meyer who were involved in patient care and provided critical review of the manuscript.

Financial support. This report did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Conflicts of interest. None.

References

1. Schiavone M, Gobbi C, Gasperetti A, et al. Congenital coronary artery anomalies and sudden cardiac death. Pediatr Cardiol 2021; 42: 1676–1687.
2. Bigler MR, Ashraf A, Seiler C, et al. Hemodynamic relevance of anomalous coronary arteries originating from the opposite sinus of Valsalva-In search of the evidence. Front Cardiovasc Med 2021; 7: 7. DOI 10.3389/fcvm.2020.591326.
3. Lee H-J, Hong YJ, Kim HY, et al. Anomalous origin of the right coronary artery from the left coronary sinus with an interarterial course: subtypes and clinical importance. Radiology 2012; 262: 101–108.
4. Baumgartner H, De Backer J, Babu-Narayan SV, et al. 2020 ESC guidelines for the management of adult congenital heart disease. Eur Heart J 2021; 42: 563–645.
5. Kuethe F, Sigusch HH, Hilbig K, et al. Detection of viral genome in the myocardium: lack of prognostic and functional relevance in patients with acute dilated cardiomyopathy. Am Heart J 2007; 153: 850–858.

https://doi.org/10.1017/S1047951122002967 Published online by Cambridge University Press