

# Bilateral Coronary Artery – Pulmonary Artery Fistula with Recurrent Ventricular Tachycardia: Case Report

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## SUMMARY

**Introduction** Bilateral coronary artery fistulae to pulmonary artery with ventricular tachycardia have not yet been described in the literature.

**Case Outline** A case of a 23-year-old male patient who was treated at our clinic for recurrent ventricular tachycardia is presented. The patient was born with six fingers on his left hand, which was surgically corrected in his early childhood. Perfusion scintigraphy demonstrated reversible ischemia at the irrigation zone of the right coronary artery. The coronary angiography revealed two coronary to pulmonary artery fistulae. The right coronary artery fistula drained through a tubular vessel formation into the pulmonary artery, but the left anterior descending fistula drained via multiple thin tortuous vessels into the pulmonary artery. The right coronary artery fistula was ligated surgically. The control scintigraphy registered no perfusion defect subsequently, but during the procedure ventricular tachycardia occurred. An electrophysiology study followed, but ventricular tachycardia could not be provoked. Two months later ventricular tachycardia occurred again. Two subsequent electrophysiology studies showed no ventricular tachycardia. The patient was treated with an implantable cardioverter defibrillator. Ventricular tachycardia was terminated four times during the first year follow-up.

**Conclusion** The mechanism of the ventricular tachycardia was unclear. The electrophysiology study was not sufficiently reliable in the patient with recurrent ventricular tachycardia and bilateral coronary artery to pulmonary artery fistulae. The therapy of choice and the prevention of sudden death in this case was an implantable cardioverter defibrillator.

**Keywords:** ventricular tachycardia; bilateral coronary artery to pulmonary artery fistulae; electrophysiology study; implantable cardioverter defibrillator

## INTRODUCTION

Congenital coronary artery fistula (CAF) is a rare malformation, which accounts for less than 1% of all cases of congenital heart diseases [1]. The incidence in the adult population is in the range of 0.1–1 [2]. CAF originates slightly more often from the right than the left coronary artery, but only 5% of them are dual [3]. Coronary artery to pulmonary artery fistula (CAPF) is very rare, accounting for only 20% of all CAFs [2].

The main clinical manifestations of CAF are coronary “steal phenomenon,” shunts at the level of the fistulas, heart failure, chest pain, infective endocarditis, arrhythmia, aneurysmal changes or fistula rupture [1, 4]. Most patients with congenital CAF remain free of symptoms [5, 6, 7].

Ventricular tachycardia (VT) complicating this infrequent congenital heart disease was randomly reported [8].

## CASE REPORT

We report the case of a 23-year-old male patient who was treated at the Institute of Cardiovascular Diseases of Vojvodina for recurrent VT.

Relevant data from anamnesis include the fact that he was born with six fingers on his left hand, which was surgically corrected in his early childhood. According to more recent medical history, the first VT was registered and treated with DC shock during a sports activity, while the second VT occurred while he was taking his examination at school.

His baseline electrocardiography showed sinus rhythm, microvoltage, with average heart rate of 80 beats per minute and signs of right heart overload (Figure 1). The echocardiographic finding was within normal values, the left ventricular ejection fraction (LVEF) was preserved, without wall motion disturbances, the stress test was without pathological findings.

The physical finding was unremarkable, except for mild systolic murmur at the base of the heart, and laboratory values were within normal ranges. The Holter electrocardiogram monitoring registered rare uniform ventricular premature beats.

The perfusion scintigraphy showed a reversible ischemia at the irrigation zone of the right coronary artery (RCA) (Figure 2).

Coronary angiography showed otherwise normal arteries, without atherosclerotic lesion, with RCA domination, but two CAPFs were found.

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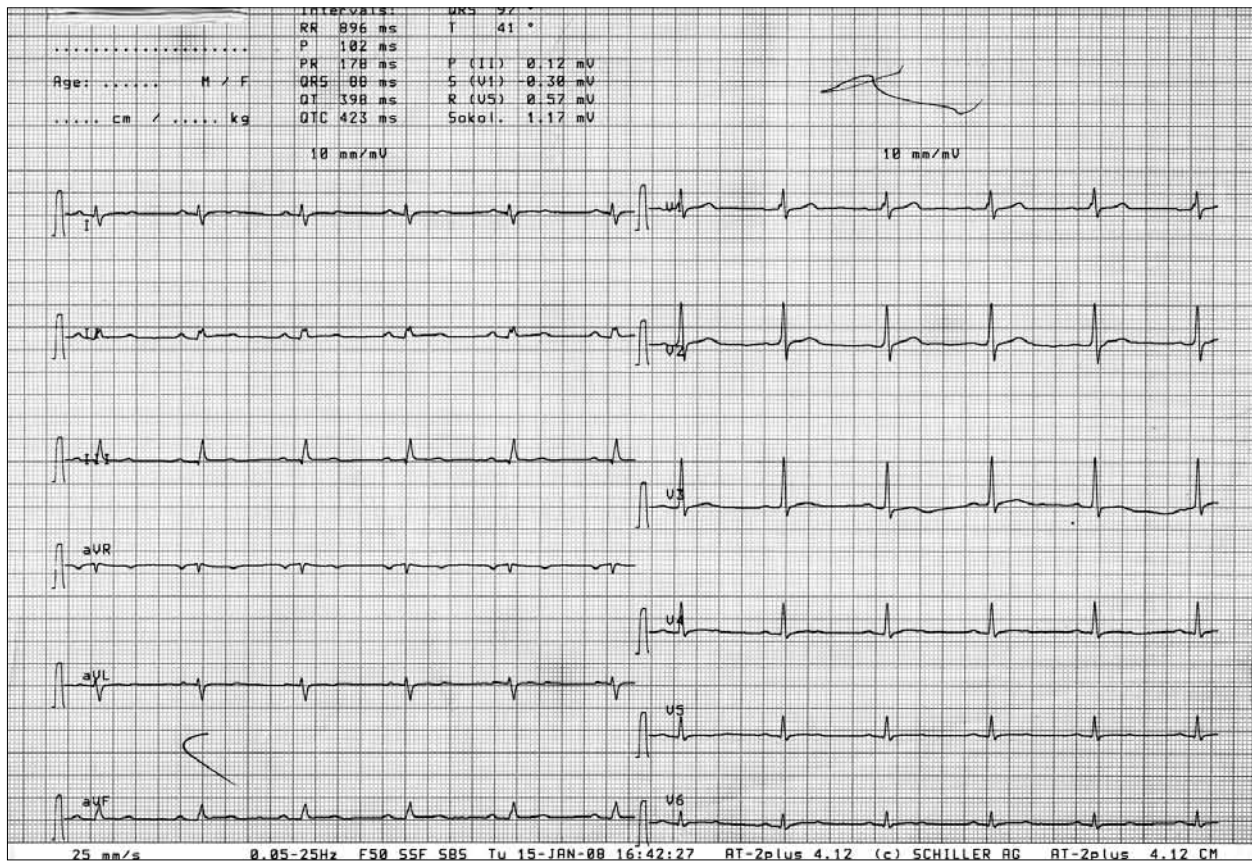


Figure 1. ECG of the patient: sinus rhythm, microvoltage, average HR about 65 bpm, right heart overload, QT interval is 398 ms, QTc is 423 ms

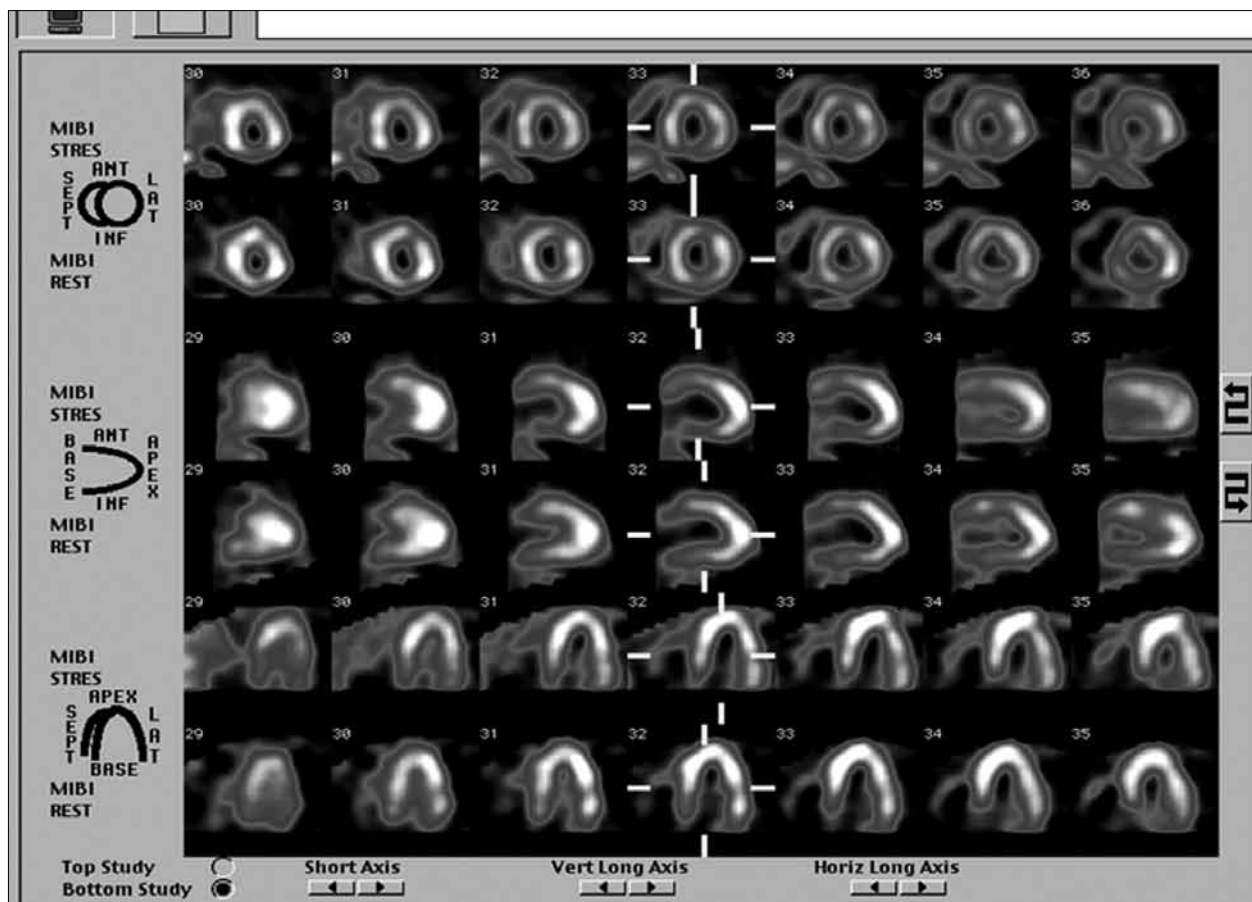


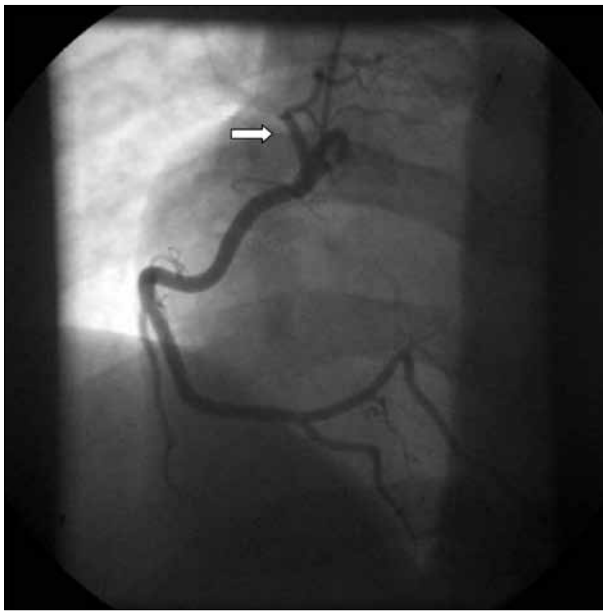
Figure 2. Reversible ischemia finding at the irrigation zone of the right coronary artery on perfusion scintigraphy

The fistula, which originated at the proximal part of the RCA, revealed patent tubular fistula emptying to pulmonary artery (Figure 3).

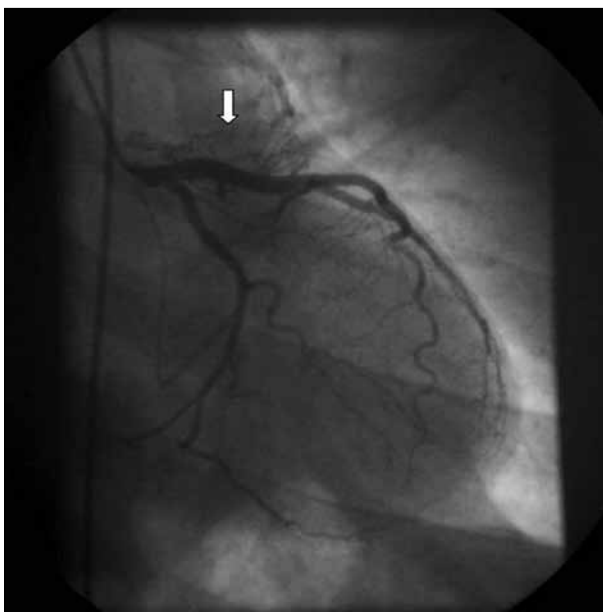
The other fistula was consisted of small thin tortuous vessels in its entire course, arising from the proximal left anterior descending artery. After a contrast injection there was faint evidence of filling the main pulmonary artery (Figure 4).

There was no evidence of shunt during heart catheterization.

The CAPF from the RCA was operatively ligated. The surgery went without any complications. Postoperatively, the patient was stable, without registered rhythm distur-



**Figure 3.** Coronary angiography of the right coronary artery in the left anterior oblique position reveals the patent tubular fistulae to the pulmonary artery



**Figure 4.** Coronary angiography of the left coronary artery in the right anterior oblique position. Fistula formed of a small thin tortuous vessels arising from the proximal left anterior descending artery and shows faint evidence of filling the main pulmonary artery

bances. The postoperative scintigraphy showed no subsequent perfusion defect (Figure 5), but during the procedure VT occurred, which had to be terminated with a DC shock. An electrophysiology study followed, but VT could not be provoked. The patient was discharged from the hospital with  $\beta$ -blockers.

Two months later, he was admitted to our clinic again because of recurrent VT, which proved to be uninducible during the electrophysiology study.

The second opinion was given by an expert in this field, who agreed with the  $\beta$ -blocker therapy. Two months later, the patient was once again brought to our clinic after resuscitation due to recurrent VT. Then, he was treated with an implantable cardioverter defibrillator. During the first year after the implantation, VT was registered and terminated successfully four times with five- and 10-joule charges.

The mechanism of VT remained unclear. In this patient, with recurrent VT and bilateral CAPF with polydactyly, the repeated electrophysiology study was not sufficiently reliable. The therapy of choice and the prevention of sudden death in this case was an implantable cardioverter defibrillator.

## DISCUSSION

Coronary artery fistula was first reported by Krause [7] in 1865. Congenital CAFs are the most common type of hemodynamically significant congenital coronary anomaly. Dual CAFs are far less common and their incidence is estimated to be around 5% in patients with this anomaly.

The right side of the heart is the drainage site in more than 90% of the cases of a single CAF.

The main pulmonary artery is the site of drainage in more than 50% of dual CAFs, as was the case in our patient. Most patients with CAF are asymptomatic and it is often an incidental finding during a coronary angiogram [8, 9, 10].

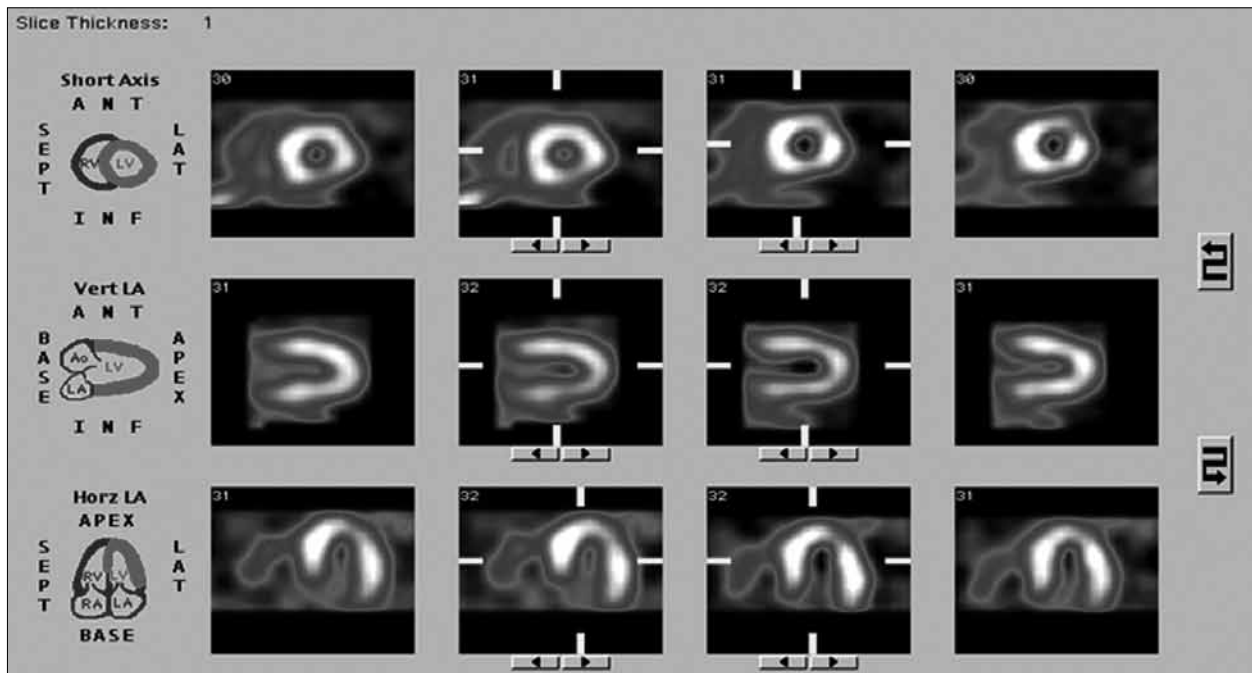
After the coronary angiography was done, the inferior wall reperfusion abnormality and the recurrent VT was explained with "steal phenomenon," due to the tubular fistula running from the RCA.

In previous papers, there were registered myocardial perfusion defects with myocardial perfusion imaging method in patients with CAPF [11], but there were also other cases, where scintigraphy failed to reveal myocardial perfusion defect even with multiple coronary-pulmonary fistulas [12] as it did in our patient at the irrigation zone of the left coronary artery. The indication for the surgical closure in our patient was clear and technically feasible, because of the registered reversible ischemia on the inferior wall and for the tubular anatomic feature of the RCA fistula.

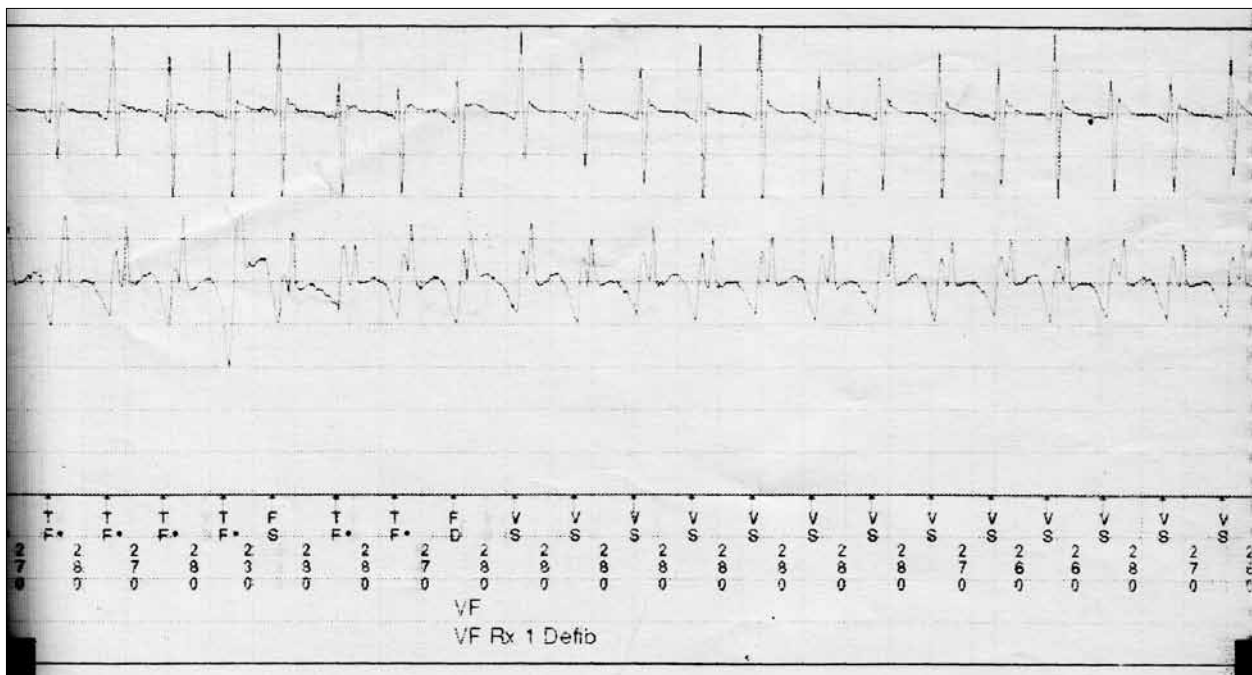
Surgical vessel ligation was first reported in 1947, and this technique remained the most frequent treatment option of CAPF [13]. This treatment modality is acceptable if the fistula is a one lumen vessel, as it was in our case.

There are data in the literature on CAPF stating that those which originate from the left anterior descending





**Figure 5.** Postoperative scintigraphy, normal finding, without perfusion defects



**Figure 6.** Strip with VT from the implanted cardioverter defibrillator report

artery with poor anatomic definition and multiple narrow tortuous tiny vessels have unclear pathogenesis and unrecognized clinical relevance [6, 9].

Treatment options for CAPF are controversial. Spontaneous occlusions are reported in early childhood, but successful cardiac surgery ligations and minimal invasive catheter techniques as embolization and occluder devices have been used as reported earlier [14–17].

Polydactyly should always arouse suspicion for various heart diseases, and its appearance can be associated with heart–hand syndromes like Holt–Oram. Once the diagnosis is made, a detailed examination and genetic counseling

is advised for other members of the patient’s family. In this case, the genetic analysis has not been done. According to our knowledge, polydactyly associated with dual CAPF and VT has not yet been listed in cardiac syndromes [18, 19].

The association of VT and preserved LVEF in individuals is well known for more than 50 years [20]. Such rhythm disturbances are usually found in young patients where, even untreated, the outcome is usually good [21, 22].

With normal morphology of the heart and preserved LVEF, VT has a tendency to be benign in its course, even if it is not treated, and it is not uncommon that provocation fails during programmed electrostimulation. According to

the Sudden Death Committee of the American Heart Association, 19% of deaths in athletes are related to coronary artery anomalies [23].

Diagnostic algorithm including physical stress test or isoproterenol test is suggested for these patients with VT [21, 22].

In patients with recurrent VT and nonischemic heart disease, nonsurgical modalities as transcatheter ablation or cryoablation could be useful, with a high rate of success [23-26].

The recurrence of noninducible VT at the electrophysiology after the surgery, even without registered ischemia at the scintigraphic finding indicated the implantation of a cardioverter defibrillator, as it is recommended by the Guidelines 1A [27].

This proved to be the therapy of choice, as during the first year after the implantation VT was terminated four times with five- and 10-joule charges (Figure 6).

During the short uneventful period after the surgery we assumed that ischemia, i.e. "steal phenomenon," was the underlying cause of VT, which was solved by surgery. The occurrence of VT after the ligation of the right-sided CAPF, which we thought to be the cause of the detectable ischemia, is unclear.

The mechanism and the cause of VT remained unclear. The electrophysiology study was not sufficiently reliable in our patient with recurrent VT and bilateral CAPF, born with polydactyly. The therapy of choice and the prevention of sudden death was an implantable cardioverter defibrillator.

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## Билатерална коронарноартеријско-плућноартеријска фистула с рецидивантном вентрикуларном тахикардијом

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### КРАТАК САДРЖАЈ

**Увод** Билатерална коронарноартеријско-плућноартеријска фистула с вентрикуларном тахикардијом није описана у литератури.

**Приказ болесника** Приказан је двадесеттворогодишњи мушкарац који је лечен због рецидивантне вентрикуларне тахикардије. Рођен је са шест прстију на левој руци, што је хируршки кориговано у раном детињству. Перфузионом сцинтиграфијом миокарда уочена је зона реверзибилне исхемије у подручју иригације десне коронарне артерије. Коронарографски је доказано постојање две коронарно-артеријско-плућноартеријске фистуле. Из десне коронарне артерије се преко тубуларне формације, а из предње нисходне артерије преко танких извијуганих крвних судова дренирала крв у плућну артерију. Урађена је хируршка лигатура фистуле десне коронарне артерије. Контролном перфузионом сцинтиграфијом се претходно описана зона исхемије више није бележила, али се током саме процедуре

јављала вентрикуларна тахикардија. Електрофизиолошким испитивањем тахикардија није провоцирана. Два месеца након тога вентрикуларна тахикардија се поновила. Две наредне електрофизиолошке студије нису провоцирале тахикардију, па је уследила уградња кардиовертер-дефибрилатора. У првој години клиничког праћења након уградње дефибрилатора вентрикуларна тахикардија је успешно обустављена у четири наврата.

**Закључак** Механизам вентрикуларне тахикардије је остао нејасан. Електрофизиолошко испитивање се показало недовољно поузданим код болесника са билатералном коронарно-артеријско-плућноартеријском фистулом. Имплантација дефибрилатора је у овом случају терапијски избор превенције изненадне срчане смрти.

**Кључне речи:** вентрикуларна тахикардија; билатерална коронарноартеријско-плућноартеријска фистула; електрофизиолошко испитивање; имплантабилни кардиовертер-дефибрилатор

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