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**Conflict of interest**

The authors declare that they have no conflicts of interest

**Keywords**

coronary fistula, vascular anomaly, congenital defect

# CORONARY PULMONARY FISTULA

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## Abstract

Coronary arteriovenous fistula (CAF) is a rare form of congenital heart disease. However, it is the most common type of congenital anomaly of the coronary arteries. [1] When there is the connection between the coronary artery and the chambers of the heart, it is called a coronary chamber fistula. The fistula may also be between a coronary artery and another adjacent vessel from the pulmonary or systemic circulation. An open fistula provides low-resistance flow by directing blood from an artery into a vein, heart chamber, or other low-pressure vessel such as the pulmonary artery. Patients with CAF may develop symptoms at birth or later in life, depending on the type of fistula and the presence of collateral circulation. Studies have reported an association between ventricular arrhythmias and sudden cardiac death syndromes in young adults and athletes with certain types of coronary anomalies, such as anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) [2-5]. The most common symptom is a myocardial ischemia. The purpose of this article is to present a clinical case of endovascular treatment of coronary pulmonary fistula. As a result of a modern diagnostic methods, such as CT angiography with three-dimensional reconstruction, it is not difficult to assess the degree and nature of the pathology. Having assessed the tactics, modern doctors are able to cope with coronary arteriovenous fistulas with great success using minimally invasive X-ray endovascular technologies.

## Коронарно-пульмональная фистула

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## Аннотация

Коронарная артериовенозная фистула (КАФ) является редкой формой врожденного порока сердца. Тем не менее, это наиболее распространенный тип врожденных аномалий коронарных артерий. Когда он находится между коронарной артерией и камерами сердца, он называется коронарно-камерным свищом. Свищ также может быть между коронарной артерией и другим соседним сосудом из легочного или большого круга кровообращения. Открытая фистула обеспечивает поток с низким сопротивлением, направляя кровь непосредственно из артерии в вену, сердечную камеру или другой сосуд с низким давлением, такой как легочная артерия. У пациентов с КАФ симптомы могут развиваться при рождении или в более позднем возрасте, в зависимости от типа фистулы и наличия коллатерального кровообращения. В исследованиях сообщалось о связи между желудочковыми аритмиями и синдромами внезапной сердечной смерти у молодых людей и спортсменов при определенных типах коронарных аномалий, таких как аномальное отхождение левой коронарной артерии от легочной артерии (ALCAPA). Одышка при физической нагрузке и стенокардия из-за ишемии миокарда являются самыми частыми симптомами. В данной статье представлен клинический случай эндоваскулярного лечения коронарно-пульмональной фистулы. На данный момент, благодаря современным методам диагностики, таких как КТ ангиография, с трехмерной реконструкцией, оценить степень и характер патологии не составляет труда. Оценив тактику, современные врачи с большим успехом способны справиться с коронарными артериовенозными фистулами с использованием малоинвазивных рентгенэндоваскулярных технологий.

**Конфликт интересов**

Авторы заявляют об отсутствии конфликта интересов

**Ключевые слова**

коронарная фистула, аномалия сосудов, врожденный порок

## Коронарлық өкпе фистуласы

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### Аңдатпа

Коронарлық артериовенозды фистула (КАФ) туа біткен жүрек ақауының сирек түрі болып табылады. Дегенмен, бұл коронарлық артериялардың туа біткен аномалиясының ең көп таралған түрі. Коронарлық артерия мен жүрек камералары арасында болса, оны коронарлық-камералық фистуласы деп атайды. Сондай-ақ, фистула коронарлық артерия мен өкпе немесе жүйелі қан айналымынан басқа іргелес тамыр арасында болуы мүмкін. Ашық фистула қанды артериядан тікелей тамырға, жүрек камерасына немесе өкпе артериясы сияқты басқа төмен қысымды тамырға бағыттау арқылы төмен қарсылық ағынын қамтамасыз етеді. КАФ бар емделушілерде фистула түріне және коллатеральды қан айналымының болуына байланысты туылғанда немесе кейінгі өмірде сипаттамалар дамуы мүмкін. Зерттеулер өкпе артериясынан сол жақ коронарлық артерияның аномалиялық шығу тегі (ALCAPA) сияқты коронарлық аномалиялардың белгілі бір түрлері бар жас ересектер мен спортшылардағы қарыншалық аритмиялар мен кенеттен жүрек өлімі синдромдары арасындағы байланыстарды хабарлады. Миокард ишемиясының әсерінен күш түскендегі ентігу және стенокардия - ең жиі кездесетін белгілер. Бұл мақалада коронарлық өкпе фистуласының эндоваскулярлық емінің клиникалық жағдайы берілген. Қазіргі уақытта үш өлшемді реконструкциямен КТ ангиографиясы сияқты заманауи диагностикалық әдістердің арқасында патологияның дәрежесі мен сипатын бағалау қиын емес. Тактиканы бағалай отырып, заманауи дәрігерлер минималды инвазивті рентгендік эндоваскулярлық технологияларды қолдана отырып, коронарлық артериовенозды фистулаларды үлкен табыспен жеңе алады.

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коронарлық фистула, қан тамырлары аномалиясы, туа біткен ақау

### Relevance

A coronary artery fistula or coronary arteriovenous fistula is a congenital or acquired defect characterized by an abnormal connection of a coronary artery with a pulmonary or systemic circulation, as well as with one or more chambers of the heart, bypassing the capillary blood flow. Coronary arteriovenous fistulas occur in the population in 0.002% of cases, and in 0.25% of patients undergoing coronary angiography [6–9].

CAF was first described by Krause in 1865. [10], revealed by coronary angiography. Haller and Little described the characteristic clinical triad of CAF: murmurs, left-to-right atrial or ventricular shunt, and dilated, tortuous coronary arteries. CAF can lead to severe hemodynamic disorders such as myocardial ischemia, cardiac arrhythmias, heart failure, and infective endocarditis in adults [11]. The classification according to Sakakibara (12) is often used to classify CAF, according to which the division according to the type of vessel origin is important: from the proximal or distal segment of the coronary arteries.

The proximal type of fistula discharge is characterized by normal diameters of the involved coronary arteries. In the case of the distal type, the entire coronary artery often expands and often ends in the right heart.

The main pathogenetic mechanism is related to the drainage of blood from high pressure blood vessels into a low pressure system through a fistula.

Causes of CAF can be congenital or acquired. More than 90% of CAFs are congenital [13]. During

early fetal development, the sinusoids feed the primitive myocardium, which is associated with the primitive tubular heart. Later, in adulthood, sinusoids usually transform into Thebesian vessels and capillaries. Persistent sinusoids that do not regress may contribute to a fistulous connection between coronary arteries and heart chambers [14, 15]. There is also a residual primitive connection between the coronary arteries and other mediastinal vessels (eg, bronchial, pericardial, or mediastinal arteries) or the superior vena cava, which may contribute to the development of a coronary arteriovenous fistula [16]. Acquired CAF results from iatrogenic events such as coronary stenting, coronary artery bypass grafting, trauma, and chest radiation [17, 18]. Some diseases, such as coronary vasculitis and myocardial infarction, can lead to the development of CHF in the chronic phase [19].

### Description of the clinical case

The purpose of this article is to present a clinical case of endovascular treatment of coronary pulmonary fistula. Patient M., 60 years old, a resident of the Almaty region, was admitted to JSC «NSC of surgery named after Syzganov A.N.» with a diagnosis of Congenital heart disease. Coronary pulmonary fistula. Atrial fibrillation. Cryoablation in 2021. CH II FC by NYHA.

The first time patient was admitted to our center in September 2021. with complaints related to cardiac arrhythmias, interruptions in the work of the heart, the patient was additionally examined at the center. Due to the AF Cryoablation was performed. Selective polypositional coronary angiography re-

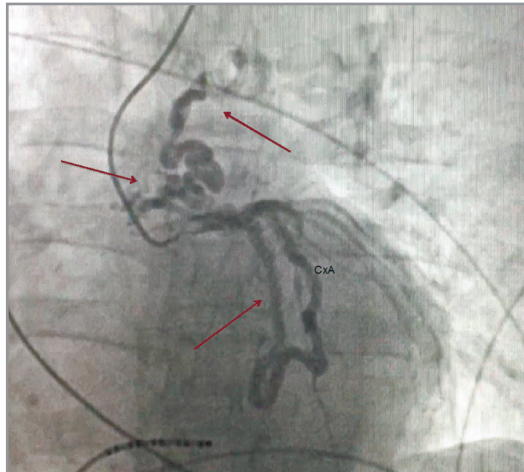
vealed anomalies associated with the coronary arteries. Because of the low ejection fraction by echocardiography (29%), was made a decision to implant a cardioverter defibrillator. The patient was discharged with an improvement in his general condition, and increasing in EF up to 36% was noted during the control EchoCG.

In January 2022 the patient is re-admitted to the Department of Interventional Cardiology, Arrhythmology and Endovascular Surgery of our Center. According to EchoCG, LV EDV is noted: 151 ml, LV ESV: 74 ml, SV 77 ml, EF 51%. interventricular septum thickness 1.2 cm. RVSP 24 mm Hg. The valves are intact. According to the ECG: Horizontal position of the electrical axis. LV hypertrophy. Myocardial ischemia of the posterior wall of the left ventricle. The patient complains of shortness of breath, mild angina pectoris, and general weakness. The patient was taken to the «cath lab»: after processing the surgical access was secured through the right radial artery using the Seldinger method. Selective polypositional coronary angiography revealed: LCA trunk - stenosis up to 30%, passable. LAD - stenosis in the proximal third up to 80%. There is an aberrant vessel in the CxA, which has a tortuous course with angiomatous nodes with a transition to an aneurysmal expansion in the final segment up to

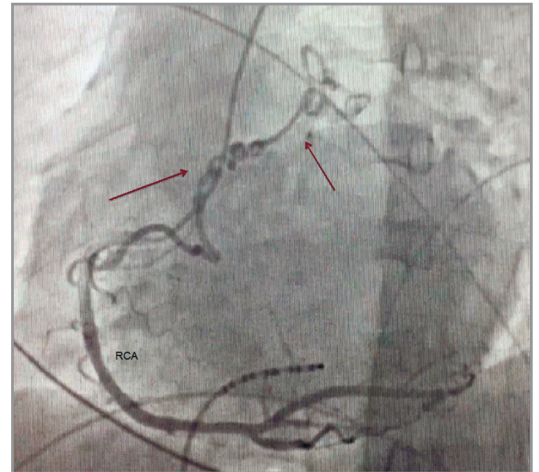
3.0 mm and a fistula into the left pulmonary artery, the diameter of the fistula is 4.5-5.0 mm. C type by Sakakibara. RCA - in the proximal third there is an aberrant vessel with a tortuous course, expanded to 5.0 mm in the final segment and a fistula with a diameter of 4.5 mm communicating with the pulmonary artery trunk. A coronary guidewire was inserted into the coronary pulmonary fistula, into it is supply vessel, on which a microcatheter «Maestro Merit» was delivered. A Nester Embolization Coil was passed through the microcatheter and delivered. During embolization, the number of coils was not enough, and therefore it was decided to use a coronary stent Ultimaster 4.5-15 mm, which was implanted in the distal third of the OA at the site of the coronary-pulmonary fistula. During control coronary angiography, the discharge of contrasted blood through the coronary-pulmonary fistula was eliminated and completely occluded.

There were no complications during the operation. The patient was transferred to the ward for dynamic observation. The patient was discharged a few days later. Subsequently, the patient is scheduled to undergo embolization of the RCA fistula and stenting of the LAD of the LCA. As a result of treatment, the patient's general condition improved, chest pains ceased to bother, shortness of breath decreased.

**Figure 1.**  
Selective coronary angiography of the LCA, direct projection. The arrows indicate tortuous aberrant vessels originating from the CxA

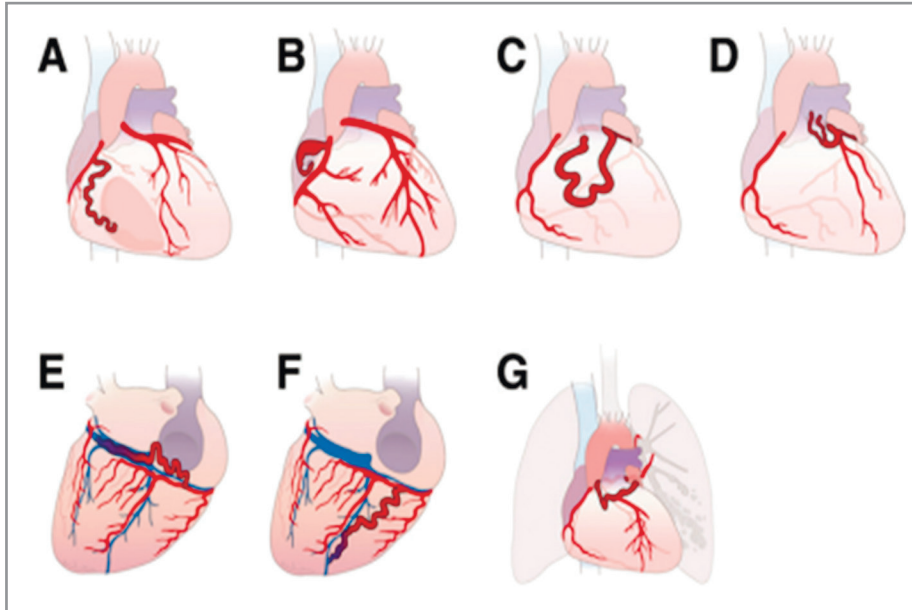


**Figure 2.**  
Selective coronary angiography of the RCA, direct projection. Arrows indicate tortuous aberrant vessels originating from the RCA CxA



**Figure 3.**  
Selective coronary angiography of the CxA, direct projection. Final view after embolization of the aberrant vessels



**Figure 4.**

Classification of CAF according to the type of drainage [12]  
 A - fistula with communication of the chamber of the right ventricle; B — fistula involving the right atrial chamber; C - fistula from the coronary artery to the pulmonary artery, with the presence of one large fistulous tract; D - coronary artery - pulmonary artery, fistula with multiple small fistulas; E - coronary artery fistula communicates with the coronary sinus; F - coronary artery-venous fistula; G - coronary artery-fistula of the bronchial artery.

### Discussions

Unfortunately, at the moment there are no single accepted protocols for the postoperative management of patients with this pathology. According to some authors, 10% of cases of transcatheter or surgical treatment were subject to CAF recanalization [20].

Zhang and colleagues [21] reported that CAF recanalization occurs within 1 year, suggesting the importance of assessing the residual shunt in the early postoperative period. Although a residual shedding of less than 2–3 mm can be observed without further intervention, careful monitoring during the postoperative follow-up period is necessary [21, 22].

Other possible complications after coil embolization or surgical ligation of the coronary arteries include persistent dilatation or aneurysmal changes in the ostia and coronary arteries, as well as the formation of blood clots, which can lead to myocardial ischemia and even infarction [23].

According to some authors, ECG changes and arrhythmias may also occur during and after

transcatheter closure [24]. In CAF with a large fistulous tract, coil migration is possible [25]. On the other hand, in patients with small branching vessels, incomplete shunt occlusion may occur if coils are placed distal to the branching point [26]. Incomplete CAF occlusion or the presence of a foreign body increases the risk of infective endocarditis [17].

### Conclusion

CAF is one of the rare pathologies with a high variability of clinical manifestations, which include such formidable complications as myocardial infarction and heart failure. As a result of a modern diagnostic methods, such as CT angiography with three-dimensional reconstruction, it is not difficult to assess the degree and nature of the pathology. Having assessed the tactics, modern doctors are able to cope with coronary arteriovenous fistulas with great success using minimally invasive X-ray endovascular technologies.

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