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Case Report

Left Circumflex Coronary Artery-to-Right Pulmonary Artery Fistula Presenting as Recurrent Angina Despite Negative Dobutamine Stress Echocardiography: A Case Report

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Abstract

Introduction: Coronary artery fistulas are abnormal communications between a coronary artery and a cardiac chamber or great vessel. Fistulas connecting the left circumflex coronary artery to the pulmonary artery are uncommon, and their clinical relevance requires assessment of both anatomical and physiological characteristics. **Case presentation:** We report the case of a 62-year-old woman with hypertension, dyslipidemia, and first-degree atrioventricular block who presented with recurrent exertional angina. Examination revealed an early diastolic murmur at the aortic area and a prominent midsystolic murmur with a palpable thrill radiating toward the right lung base. Echocardiography showed moderate aortic regurgitation with preserved left ventricular systolic function, while dobutamine stress echocardiography showed no inducible wall-motion abnormalities. She subsequently developed recurrent angina with new T-wave inversions and mildly elevated high-sensitivity cardiac troponin T concentrations that remained unchanged across serial measurements. Coronary angiography demonstrated nonobstructive epicardial coronary arteries and a fistula arising from the atrioventricular nodal branch of the distal left circumflex artery and draining into the right pulmonary artery. Coronary steal was suspected but not physiologically confirmed. Following individualized assessment, the Heart Team recommended closure; however, the patient declined intervention and remained free of recurrent angina during approximately five months of follow-up. **Conclusions:** This case highlights the distinction between an anatomically confirmed coronary artery fistula and its unconfirmed physiological significance. Negative stress echocardiography and alternative potential contributors to symptoms should be considered when interpreting suspected coronary steal. Management requires individualized multidisciplinary assessment and shared decision-making.

Keywords: coronary artery fistula; coronary artery-to-pulmonary artery fistula; left circumflex artery; coronary steal; recurrent angina; dobutamine stress echocardiography; nonobstructive coronary arteries

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1. Introduction

Coronary artery fistulas (CAFs) are rare congenital or acquired coronary anomalies characterized by abnormal communications between a coronary artery and a cardiac chamber or great vessel. They are typically identified incidentally during invasive coronary angiography or coronary computed tomography angiography, with a reported incidence of approximately 0.1–0.2% among patients undergoing diagnostic coronary angiography. The right coronary artery is the most common site of origin, accounting for approximately 50–60% of cases, followed by the left anterior descending artery (25–42%). The majority of fistulas drain into low-pressure right-sided cardiac structures, most frequently the right ventricle, right atrium, or pulmonary artery, in descending order of frequency [1,2].

Most CAFs remain asymptomatic, particularly when small. Clinically significant fistulas may be associated with myocardial ischemia, arrhythmias, or heart failure. Potential mechanisms include coronary steal and volume overload, depending on fistula anatomy and shunt magnitude; however, their contribution requires patient-specific assessment [2,3].

We report the case of a woman with hypertension, dyslipidemia, and first-degree atrioventricular block who presented with recurrent angina. Coronary angiography identified a fistulous communication arising from the atrioventricular nodal branch of the distal left circumflex coronary artery and draining into the right pulmonary artery. Given the recurrent symptoms and dynamic electrocardiographic changes, the lesion was considered potentially clinically significant, with coronary steal regarded as a possible, but unconfirmed, mechanism.

2. Case Presentation

A 62-year-old woman with a 15-year history of dyslipidemia (treated with atorvastatin 80 mg once daily), arterial hypertension (treated with amlodipine 5 mg once daily and losartan 100 mg once daily), and first-degree atrioventricular block documented on prior electrocardiogram was evaluated for recurrent chest pain.

Her surgical history included cholecystectomy, right inguinal herniorrhaphy, and cystopexy. Family history was notable for bicuspid aortic valve in her son and grandson, without other known cardiovascular disease in first-degree relatives. The patient's clinical course is summarized in Figure 1.

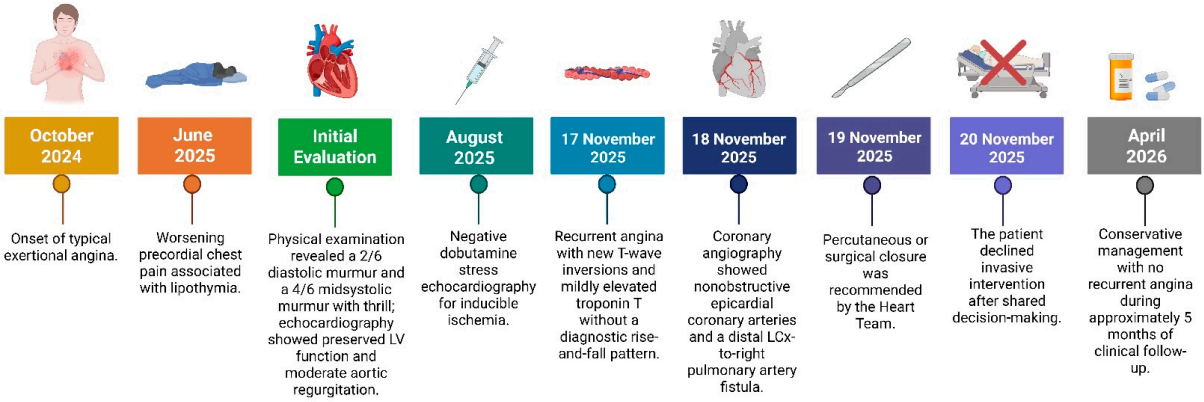


Figure 1. Clinical timeline of the case.

The patient reported typical exertional angina beginning in October 2024. In June 2025, she experienced a new episode of more intense chest pain associated with lipothymia. On physical examination, blood pressure was 130/70 mmHg, heart rate was 79 bpm, respiratory rate was 20 breaths per minute, and oxygen saturation was 94% on room air. Cardiac auscultation revealed a regular rhythm with a grade 2/6 early diastolic decrescendo murmur

best heard at the aortic area, and a grade 4/6 midsystolic murmur with a palpable thrill radiating to the right lung.

Transthoracic echocardiography demonstrated preserved global and regional left ventricular systolic function, with a left ventricular ejection fraction of 64% by Simpson's biplane method and a global longitudinal strain of −23%, without regional wall-motion abnormalities. Left ventricular end-diastolic and end-systolic diameters were 4.0 and 2.5 cm, respectively, with an indexed left ventricular mass of 62.69 g/m^2. Left atrial area was 16.3 cm^2, estimated pulmonary artery systolic pressure was 24 mmHg, and the E/e′ ratio was 9.16. Indexed left ventricular volumes and left atrial volume index were not available in the records reviewed for this report. Aortic regurgitation was graded as moderate, with a vena contracta width of 5 mm and a pressure half-time of 385 ms. An eccentric regurgitant jet extended to the mid-left ventricle, and no holodiastolic flow reversal was observed in the descending aorta. These findings showed no left ventricular dilatation or echocardiographic evidence of elevated filling pressures at the time of assessment. Nevertheless, moderate aortic regurgitation remained a potential contributor to the elevated NT-proBNP concentration and clinical presentation. The additional grade 4/6 midsystolic murmur with a palpable thrill prompted further investigation for a concomitant structural lesion.

Given persistent symptoms and suspected coronary artery disease, dobutamine stress echocardiography was performed using a maximum dobutamine infusion rate of 50 µg/kg/min without atropine. The patient continued her usual cardiovascular medications, including amlodipine and losartan, without withholding them before the examination. Heart rate increased from 74 beats/min at rest to 136 beats/min at peak stress. The examination was terminated after the target heart rate and maximum protocol dose were reached. No inducible regional wall-motion abnormalities, anginal symptoms, or ischemic electrocardiographic changes were observed, including at the maximum dobutamine dose.

Despite the negative stress test, the patient subsequently presented with recurrent chest pain with typical anginal characteristics and new T-wave inversions. Laboratory findings are summarized in Table 1. High-sensitivity cardiac troponin T was measured using a Roche Elecsys assay at presentation and at 1 and 3 h, with a reported concentration of 0.03 ng/mL at each measurement. The concentrations remained unchanged across the three measurements, without a demonstrated rise-and-fall pattern. NT-proBNP was elevated at 1658 pg/mL. Given the recurrent symptoms and new electrocardiographic changes, the Heart Team proceeded with invasive coronary angiography.

Table 1. Laboratory profile at the acute anginal evaluation.

Test	Result	Reference Range
WBC (×10^3/µL)	4.86	3.4–9.7
Neutrophils (×10^3/µL)	2.48	2.2–4.8
Lymphocytes (×10^3/µL)	1.62	1.1–3.2
Hemoglobin (g/dL)	15	12.0–16.0
Platelets (/µL)	161,000	130,000–400,000
High-sensitivity cardiac troponin T (ng/mL)	0.03	0–0.014
NT-proBNP (pg/mL)	1658	<125
Creatinine (mg/dL)	0.96	0.5–1.1
LDL cholesterol (mg/dL)	88	<100
HDL cholesterol (mg/dL)	38	40–59
Triglycerides (mg/dL)	192	<150

Reference intervals are those reported by the laboratory. The hemoglobin interval is presented as a laboratory reference interval; altitude-specific validation was not established for this report.

Coronary angiography demonstrated a right-dominant system with nonobstructive epicardial coronary arteries. A fistulous communication arose from the atrioventricular nodal branch of the distal left circumflex coronary artery and drained into the right pulmonary artery. Distal LCx opacification was preserved on angiography (Figure 2A–D). The available angiographic report did not provide quantitative fistula dimensions, a detailed description of its course, or explicit characterization of proximal coronary or aneurysmal dilatation. These findings established the anatomical diagnosis but did not quantify shunt magnitude or demonstrate reduced myocardial perfusion attributable to the fistula. Coronary steal was considered a possible explanation for the clinical presentation.

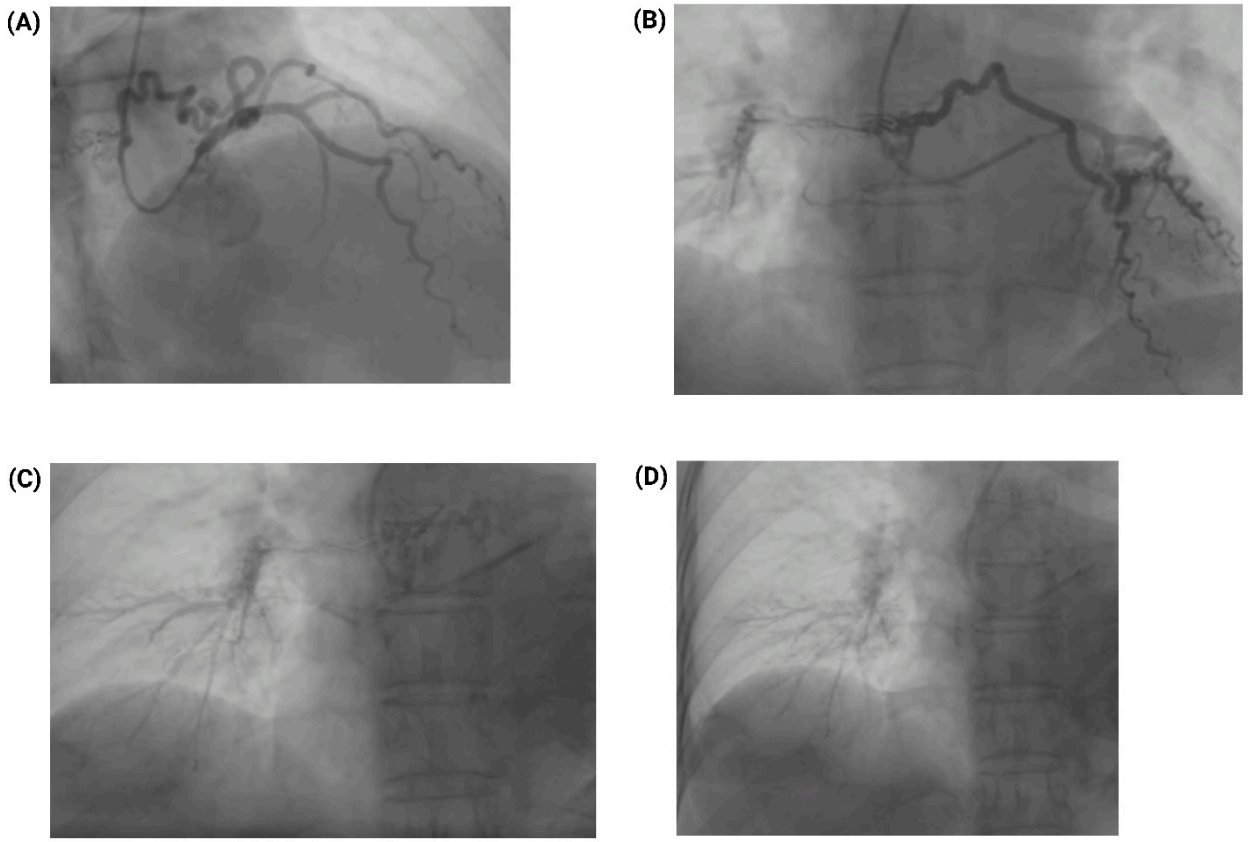


Figure 2. Invasive coronary angiography demonstrating a left circumflex coronary artery-to-right pulmonary artery fistula. (**A**,**B**) Selective left coronary angiography showing a fistulous communication arising from the atrioventricular nodal branch of the distal left circumflex coronary artery. (**C**,**D**) Delayed opacification of the right pulmonary artery through the fistulous tract. LCx, left circumflex coronary artery; RPA, right pulmonary artery.

Following individualized assessment of the recurrent symptoms, electrocardiographic changes, and angiographic findings, the multidisciplinary Heart Team recommended fistula closure, with percutaneous and surgical approaches considered. The patient declined intervention after discussion of the potential benefits, procedural risks, and uncertainty regarding the contribution of the fistula to her symptoms. Cardiovascular risk factor management was continued, and she remained free of recurrent angina during approximately five months of clinical follow-up.

3. Discussion

CAFs are rare coronary anomalies, with an incidence of 0.1–0.2% among patients undergoing coronary angiography and as low as 0.002% in the general population [4,5]. They are primarily congenital in origin, although secondary forms have been described

in the context of inflammatory diseases such as Kawasaki disease or Takayasu arteritis, as well as following intracardiac device implantation, cardiac surgery, endomyocardial biopsy, or direct chest trauma [3,6]. CAFs are characterized by an anomalous fistulous connection between a coronary artery and an adjacent cardiac chamber or great vessel, including the right ventricle, right atrium, superior vena cava, or pulmonary artery, thereby bypassing the myocardial capillary bed and creating a pathway of minimal resistance to flow [3]. The coronary artery to pulmonary artery fistula (CPAF) represents an uncommon subtype, accounting for approximately 15–30% of all CAFs, and is frequently diagnosed incidentally during coronary angiography or noninvasive cardiac imaging [5,7]. Published reports describe fistulas arising from different coronary arteries, including the LCx [5]. However, distributions derived from case reports and case series are subject to selection and publication bias and should not be interpreted as population prevalence estimates or used to establish a ranking of anatomical rarity [5].

Fistula size, anatomy, shunt magnitude, associated complications, and patient characteristics are relevant to management decisions [3,8]. These features should be evaluated individually rather than inferring physiological significance from symptoms or qualitative angiographic appearance alone. In the published cases compiled by Wu et al., the mean age at diagnosis was 56.3 years. Nonspecific symptoms were commonly reported, whereas typical angina and an auscultatory murmur were reported in 9.1% and 16.3% of cases, respectively. These descriptive findings are subject to selection and reporting bias and should not be generalized to unselected patients with CPAF [5]. The clinical presentation of CPAFs is heterogeneous and often nonspecific: more than two thirds of patients presented with nonspecific symptoms such as chest pain, dyspnea on exertion, or palpitations, while only 9.1% exhibited typical angina pectoris, and a cardiac murmur was auscultated in merely 16.3% of cases [5]. In the present case, recurrent exertional angina, dynamic T-wave changes, and the angiographically documented fistula raised suspicion of a fistula-related ischemic mechanism. Coronary steal provides a plausible explanation; however, reduced distal myocardial perfusion was not demonstrated. Accordingly, the anatomical diagnosis should be distinguished from the unconfirmed physiological mechanism, and neither the murmur nor the clinical presentation alone establishes a hemodynamically significant shunt [2,9].

A particularly relevant feature of this case is the absence of inducible wall-motion abnormalities on dobutamine stress echocardiography. In the review by Wu et al., 65 of 78 patients undergoing stress testing had positive results (83.3%); this selected subgroup of published cases should not be considered representative of all patients with CPAF [5]. The negative examination in our patient cannot be classified as false-negative because ischemia was not independently demonstrated. A limited myocardial territory or perfusion abnormalities insufficient to produce detectable wall-motion changes could potentially explain a negative examination. However, these explanations remain speculative without dedicated perfusion imaging or physiological assessment [3,5]. The examination was performed while the patient continued her usual cardiovascular medications, which should also be considered when interpreting the findings.

Invasive coronary angiography established the anatomical diagnosis in this patient, although its two-dimensional nature limits comprehensive characterization of the fistulous course and surrounding structures. Coronary computed tomography angiography can provide complementary three-dimensional information about the origin, course, drainage site, and associated aneurysmal changes, facilitating procedural planning [3,5]. Its absence in this case limited anatomical characterization. Anatomical imaging findings alone, however, do not establish the physiological significance of the fistula.

The 2025 ACC/AHA/HRS/ISACHD/SCAI guideline emphasizes assessment by an experienced multidisciplinary team to determine the role of medical therapy, transcatheter

closure, or surgery in patients with coronary artery fistulas [10]. In this patient, closure was recommended after consideration of recurrent symptoms, dynamic electrocardiographic changes, and angiographic findings. This was an individualized clinical recommendation rather than an indication established by quantified shunting or independently demonstrated fistula-related ischemia. The patient's decision to decline intervention was respected, and conservative management with continued follow-up was pursued.

Both untreated fistulas and closure procedures may be associated with adverse outcomes. Wu et al. reports eight deaths among 439 cases (1.8%): two sudden cardiac deaths, two aneurysm ruptures, one postoperative embolic stroke, two deaths following failed percutaneous interventions, and one left main dissection [5]. These heterogeneous published outcomes cannot estimate this patient's individual risk or establish a survival benefit from closure. Aneurysmal dilatation was not explicitly characterized in the available angiographic report; therefore, aneurysm-related risk could not be adequately assessed in this case.

Although the fistula provided a plausible anatomical substrate for the symptoms, alternative mechanisms of angina with nonobstructive coronary arteries, including coronary microvascular dysfunction and vasomotor abnormalities, could not be excluded, consistent with the diagnostic framework of the 2024 ESC guideline [11]. Moderate aortic regurgitation also remained a potential contributor to the clinical presentation and elevated NT-proBNP. Continued clinical follow-up and reassessment are therefore important, particularly if symptoms recur.

If intervention is reconsidered, further anatomical characterization and, where clinically appropriate, physiological assessment would help inform the decision. Selection between transcatheter and surgical closure should account for the fistula's origin, course, drainage site, relationship to myocardial branches, and procedural feasibility. Potential complications, including compromise of coronary branches, thrombosis, and myocardial infarction, should be incorporated into multidisciplinary assessment and shared decision-making [3,10]. Surgical ligation, as performed by Aydin et al. in a proximal LCx-to-pulmonary artery fistula with complete normalization of the stress test at two months postoperatively, remains a viable and definitive alternative, particularly for complex fistulas or when concomitant cardiac surgery is required [12].

4. Limitations

This case has several limitations. Coronary computed tomography angiography was not performed, limiting three-dimensional characterization of the fistula and its relationship to adjacent coronary branches. Quantitative fistula dimensions, a detailed description of its course, and explicit characterization of proximal coronary and aneurysmal dilatation were unavailable in the angiographic report. Indexed left ventricular volumes and left atrial volume index were also unavailable. Shunt magnitude, fistula flow, and coronary physiological indices were not quantified, and dedicated myocardial perfusion imaging was not performed. Therefore, coronary steal and the physiological significance of the fistula remain unconfirmed. Although echocardiography provided an estimated pulmonary artery systolic pressure, this did not establish the magnitude of a fistula-related shunt. Serial hs-cTnT concentrations were unchanged at the reported precision, and their elevation could not be attributed specifically to the fistula. Coronary microvascular dysfunction, vasomotor abnormalities, and the contribution of moderate aortic regurgitation could not be excluded. Finally, the absence of post-closure assessment and the approximately five-month follow-up limit causal interpretation and conclusions regarding long-term outcomes.

5. Conclusions

This case describes an angiographically documented fistula arising from the atrioventricular nodal branch of the distal left circumflex coronary artery and draining into the right pulmonary artery in a patient with recurrent angina and negative dobutamine stress echocardiography. Coronary steal was suspected but not confirmed, and alternative contributors to symptoms remained possible. The case underscores the need to distinguish anatomical findings from physiological significance and to individualize management through multidisciplinary assessment, shared decision-making, and continued follow-up.

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References

1. Hang, K.; Zhao, G.; Su, W.; Bao, G.; Zhao, Q.; Jiao, Z.; Tian, Z.; Zhang, H.; Nie, L.; Luo, R.; et al. Coronary artery-to-pulmonary artery fistula in adults: Evaluation with 320-row detector computed tomography coronary angiography. *Ann. Transl. Med.* **2021**, *9*, 1434. [CrossRef] [PubMed]
2. Hamad, L.M.B.; Al Azzoni, A.; Mubarak, W. Coronary Steal Caused by a Rare Left Circumflex–Pulmonary Artery Fistula: A Diagnostic and Therapeutic Challenge—A Case Report from Ain Al Khaleej Hospital. *Ann. Clin. Case Rep.* **2025**, *10*, 2807.
3. Al-Hijji, M.; El Sabbagh, A.; El Hajj, S.; AlKhouli, M.; El Sabawi, B.; Cabalka, A.; Miranda, W.R.; Holmes, D.R.; Rihal, C.S. Coronary Artery Fistulas. *JACC Cardiovasc. Interv.* **2021**, *14*, 1393–1406. [CrossRef] [PubMed]
4. Yamanaka, O.; Hobbs, R.E. Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. *Cathet Cardiovasc Diagn.* **1990**, *21*, 28–40. [CrossRef] [PubMed]
5. Wu, C.; Joseph, S.; Devireddy, R.; Kaldas, M.; Zlatopolsky, M.; Qaqish, O.; Gong, Y.C.; Blaceri, S.; Saad, C.; Harder, E.W.; et al. A Systematic Analysis of Coronary to Pulmonary Artery Fistula and Its Associated Aneurysm in Adults. *Interdiscip. Cardiovasc. Thorac. Surg.* **2025**, *40*, ivaf206. [CrossRef] [PubMed]
6. Stout, K.K.; Daniels, C.J.; Aboulhosn, J.A.; Bozkurt, B.; Broberg, C.S.; Colman, J.M.; Crumb, S.R.; Dearani, J.A.; Fuller, S.; Gurvitz, M.; et al. 2018 AHA/ACC Guideline for the Management of Adults with Congenital Heart Disease. *Circulation* **2019**, *139*, e698–e800. [CrossRef] [PubMed]
7. Verdini, D.; Vargas, D.; Kuo, A. Coronary-pulmonary artery fistulas: A systematic review. *J. Thorac. Imaging* **2016**, *31*, 380–390. [PubMed]
8. Saeed, A.; Ghumman, G.M.; Mir, D.; Khan, A.A.; Sivarama, K. Left Coronary Artery to Pulmonary Trunk Fistula: Two Case Reports with Literature Review. *Cureus* **2023**, *15*, e43672. [CrossRef] [PubMed]
9. Lim, E.C.; Lim, C.E. Coronary Artery Fistula Presenting with Coronary Steal Syndrome: The Significance of Clinical Vigilance and Second Opinions. *Clin. Case Rep.* **2025**, *13*, e70874. [CrossRef] [PubMed]

10. Gurvitz, M.; Krieger, E.V.; Fuller, S.; Davis, L.L.; Kittleson, M.M.; Aboulhosn, J.A.; Bradley, E.A.; Buber, J.; Daniels, C.J.; Dimopoulos, K.; et al. 2025 ACC/AHA/HRS/ISACHD/SCAI Guideline for the Management of Adults with Congenital Heart Disease: A Report of the American College of Cardiology/American Heart Association Joint Committee on Clinical Practice Guidelines. *Circulation* **2026**, *153*, e115–e251. [PubMed]
11. Vrints, C.; Andreotti, F.; Koskinas, K.C.; Rossello, X.; Adamo, M.; Ainslie, J.; Banning, A.P.; Budaj, A.; Buechel, R.R.; Chiariello, G.A.; et al. 2024 ESC Guidelines for the management of chronic coronary syndromes. *Eur. Heart J.* **2024**, *45*, 3415–3537. [CrossRef] [PubMed]
12. Aydin, S.; Altiparmak, I.H.; Yildiz, A.; Demirbag, R.; Yilmaz, R. Rare coronary artery anomaly: Left circumflex coronary artery fistula draining into the pulmonary artery. *Cardiovasc. Revascularization Med. Incl. Mol. Interv.* **2008**, *9*, 182–183. [CrossRef] [PubMed]

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