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Coronary Artery Fistula Presenting With Coronary Steal Syndrome: The Significance of Clinical Vigilance and Second Opinions

Enoch Chi Ngai Lim[1] | Chi Eung Danforn Lim[1,2,3]

[1]Translational Research Department, Specialist Medical Services Group, Sydney, New South Wales, Australia | [2]NICM Health Research Institute, Western Sydney University, Westmead, New South Wales, Australia | [3]School of Life Sciences, University of Technology Sydney, Ultimo, New South Wales, Australia

Correspondence: Chi Eung Danforn Lim (chi.lim@westernsydney.edu.au)

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ABSTRACT

Safety should always come first for the patient. A change in clinical picture should trigger clinicians to seek a fresh perspective and reassess, even if they have been reassured by other specialists, as "innocent" coronary artery fistulae can evolve into serious and potentially deadly conditions.

1 | Introduction

Coronary artery fistulae (CAFs) are abnormal communications between a coronary artery and a cardiac chamber or great vessel, bypassing the myocardial capillary bed [1]. They account for 0.8% of congenital coronary anomalies, with a prevalence of 0.002% in the general population and 0.1%–0.2% in patients undergoing catheterisation or angiography [1, 2]. No sex or ethnic predilection is recognized [1]. Most arise in isolation, though some accompany other congenital defects. Acquired cases, from trauma or interventions, are rare. Over 90% of CAFs drain into right-sided chambers or pulmonary artery, producing a left-to-right shunt [3]. Less commonly, they terminate in the left heart, leading to volume overload [2]. Haemodynamic effect depends on size, tortuosity, and drainage site [2]. Small fistulae are usually silent; large ones divert blood during diastole, causing coronary steal and ischaemia. Large right-sided fistulae may cause right heart overload and pulmonary overcirculation.

Most patients are asymptomatic when young, with detection often incidental [1]. With age, declining reserve and fistula enlargement increase clinical expression [1, 4]. By the fifth decade, symptoms are common, and over two-thirds of patients >60 years are affected [1]. Manifestations range from fatigue and dyspnoea to angina, arrhythmias, heart failure, or infarction [1, 2]. Rare complications include sudden death and endocarditis [2]. Physical findings may be absent in small CAFs, but large high-flow lesions often produce a continuous murmur [1].

Diagnosis is challenging. TTE can show dilation and turbulence but lacks spatial resolution; Doppler or contrast-enhanced TEE improves accuracy [1]. ECG is usually normal, though chronic shunts may show chamber strain [1]. Coronary angiography remains the gold standard, defining origin, drainage, and shunt effect [5–7]. Computed Tomography Coronary Angiography (CTCA) provides detailed arterial anatomy and aids surgical planning [7, 8]. CTCA and MRI angiography enhance non-invasive assessment, often revealing fistulae undetected by echocardiography [1]. Advanced tools, including Doppler wire flow quantification and computational fluid dynamics, evaluate coronary steal and risks of thrombosis or rupture [6–9]. Myocardial perfusion imaging,

such as thallium-dipyridamole scanning, demonstrates reversible ischaemia attributable to coronary steal [10, 11].

As shown in Table 1, various imaging modalities can be used to detect CAF, but coronary angiography remains the most valid method for determining diagnostic and CAF's haemodynamic significance [4]. For our patient, the fistula was first visualized on CTCA. However, the clinical importance was deemphasized due to a lack of symptoms, underscoring a diagnostic quandary: differentiating accurately benign incidental imaging findings from those which have latent pathogenic potential.

Existing research from the literature summarizes that CAFs in adults points to a lack of clear management protocols, particularly for incidentally discovered cases concerning older people. Many asymptomatic fistulae of CAD have been treated conservatively in the past because of an oversimplified belief that they are of benign nature. As this case illustrates, however, "asymptomatic" does not mean harmless. There are gaps in longitudinal studies investigating the progression over time of small, asymptomatic fistulae in adults, which creates an environment conducive to the misinterpretation or underappreciation of such anomalies. CAF was regarded as incidental by the patient's cardiologist and deemed as clinically inconsequential, considering the absence of associated symptoms. The novel aspects of this case are the overdue clinical assumption of marked symptomology, together with a need to reconsider the previous diagnosis, which may have underestimated clinical significance. This highlights the need to continue maintaining a watchful eye for rethinking and revisiting assumptions: clinicians need to be cautious that a previously benign abnormality may become pathologic and also be associated with symptoms. It also demonstrates how helpful collaborative teams, alongside diagnostic conflict, addressing nuances from single network surveillance for unusual conditions, can be safe and accurate.

TABLE 1 | Comparison of Imaging Modalities for Coronary Artery Fistula.

Imaging modality	Key features
Transthoracic echocardiogram [1]	• Can demonstrate coronary artery dilation and flow turbulence • Unable to visualize the anatomy
Coronary angiography [5, 6]	• Gold standard for diagnosing CAF • Visualizes fistulous connection and steal phenomenon
Computed tomography [7, 8]	• Provides detailed coronary architecture • Guides therapeutic interventions
Doppler wire and computational fluid dynamics [6, 9]	• Measures flow rates • Assesses hemodynamic changes and complication risks
Myocardial perfusion [10, 11]	• Demonstrates reversible ischemia caused by steal phenomenon

2 | Case History / Examination

A 68-year-old Chinese man came to see a GP for a second opinion because of concerns about feeling faint. He described multiple episodes of near fainting in the previous several weeks, with mild exertion or stress inducing these episodes, e.g., while he was singing hymns in the church during a Sunday service. These episodes included light-headedness, blurring of vision, and weakness without complete loss of consciousness. While there was no pain in the chest, palpitations, or considerable breathlessness during the episodes, he reported a decline in exercise tolerance. There were no signs of orthostatic hypotension or prodromal features of vasovagal syncope.

His medical history included well-controlled hypertension and dyslipidaemia for which he was on regular medications (ACE-inhibitor and statin). There were no significant prior myocardial infarction, arrhythmias, or structural heart disease. Previously completed evaluation in 2023 for atypical chest discomfort also included a CT coronary angiogram about 18 months before this visit. That CTCA showed a coronary artery anomaly; specifically, there was a fistulous connection from the LAD to the pulmonary trunk. Other coronary arteries did not show significant atheromatous stenosis. He was seeing another cardiologist at the time who thought the fistula was a small, incidental finding without any significant impact on blood flow. The patient was told that he did not need any intervention and his chest symptoms were treated as probable gastro-esophageal reflux. He was instructed to be seen on a routine basis in 2 years time, but there was no specific cardiac follow-up regarding the fistula itself. Until the recent presyncopal episodes, he remained mostly asymptomatic.

Worried about the new symptoms, the patient decided to visit another GP for a second opinion, since his usual GP told him to follow what the cardiologist had said. The second GP carefully examined the relevant history and promptly recalled coronary fistulas of concern in a presyncope context and urgently requested a consult with another cardiologist. At presentation, the patient looked well, with normal cognition. His blood pressure was 130/80 mmHg, and pulse showed a heart rate of 72 bpm, which was regular. Cardiovascular examination revealed normal heart sounds and no significant murmur while supine. However, on upright posture and after mild exertion (bedside step-up test), a faint high-pitched continuous murmur was found along the left sternal border. Although faint, this addition escalated the hypothesis of a significant left-to-right shunt. There were no signs of heart failure, in which the lung fields were devoid of sounds and there was no peripheral oedema. Peripheral pulses were palpable and symmetrical. Neurological exam was essentially normal without any remarkable findings, and there was no audible carotid bruits.

3 | Investigation, and Treatment

The 12-lead ECG was normal and had sinus rhythm, achieving 70 bpm. Acute ischemia was not observed, although there was mild and non-specific ST-T wave flattening in the lateral leads that was unchanged from prior recordings. A transthoracic echocardiogram revealed normal left ventricular size and systolic function. There was a left to right shunt seen at the level of the right ventricular outflow tract, consistent with the known coronary artery fistula to the pulmonary trunk identified from the CTCA in 2023. It demonstrated a complex coronary anomaly with a coronary arcade between the proximal LAD and proximal RCA, as well as a possible single fistula between the arcade and the pulmonary trunk. The cardiologist explained to the patient that his symptoms are suggestive of ischemia secondary to coronary steal syndrome, as LAD flow from the LAD was likely redirected through collateral connections to the RCA and into the pulmonary trunk, contributing to the steal phenomenon.

4 | Conclusion and Results

The cardiologist expedited a referral to a cardiothoracic surgeon, who organized an urgent coronary angiography, which revealed a considerable coronary arteriovenous fistula. A large AV fistula extending from the proximal LCA to the pulmonary artery with multiple feeding vessels was noted. The native coronary arteries appeared to be free of obstruction, but the flow in the distal LAD did seem slow, presumably due to a steal phenomenon. Coronary steal causes reduced perfusion pressure in the distal LAD as blood preferentially flows through the low-resistance fistulous connections, thereby limiting forward flow to the dependent myocardium. No plaque formation or stenosis was observed at the egress points of the fistulae, indicating that the coronary steal was due to the anatomical shunt rather than atherosclerotic disease. The right coronary was dominant. It was also widely patent, but there were AV connections from the proximal RCA to the pulmonary artery. The angiography revealed separate feeding arteries from both the RCA and LAD, confirming dual fistulae.

The patient's case was reviewed during the multidisciplinary cardiothoracic-cardiology conference of the tertiary university teaching hospital that incorporates interventional cardiologists, cardiothoracic surgeons, and radiologists. The patient was considered for coronary artery fistulae closure due to the patient's symptomatology and angiography showing substantial shunting and steal. Although Qp:Qs was not formally measured, the degree of angiographic shunting was significant and supported the clinical decision for intervention. Despite coil embolization or device implantation being considered the least invasive approach to transcatheter closure of congenital arterial fistulae, which is frequently the initial strategy in uncomplicated cases, this patient was classified as having more complex anatomy than preferred for an endovascular approach. As a result, the hospital team chose surgical intervention for complete correction. Comprehensive counseling was done for the patient, including discussion of the corrective surgery risks (sternotomy, cardiopulmonary bypass, etc.) versus the risks of no intervention (ongoing presyncope, potential for myocardial infarction or heart failure). His quality of life was impacted considerably, so he was inclined to proceed with active treatment.

5 | Outcome and Follow-Up

Under general anesthesia, coronary artery fistulae were surgically repaired via a median sternotomy that was elective in nature. Cardiopulmonary bypass was established via the aorta and right atrium. The heart was arrested with cold antegrade blood cardioplegia. Intraoperative findings confirmed that biventricular function was preserved, and there was no significant valvular pathology. Major conducting fistulous vessels were noted over the pulmonary artery, arising from both the proximal right coronary artery (RCA) and left anterior descending artery (LAD). Flow from these fistulae into pulmonary artery jets was noted from two discrete jets of flow. Correlating TOE findings preoperatively had demonstrated a communication from the LAD to the anterior wall of the pulmonary artery, and there was suspicion of a connection next to the left atrial appendage (LAA). Mobilization and tracing of the fistulous tracts were carried out to the RCA and LAD, where ligation and division were done. Two openings gained in the pulmonary artery had 6–0 Prolene used to oversew them. After 125 min, the cross clamp was removed. Total perfusion time was 142 min, with successful weaning from bypass. Patient status post bypass was stable.

Repeat TOE after the repair showed resolution of the previously visualized jets on color Doppler and no additional evidence of communication between the coronary artery or LAD and pulmonary artery. Biventricular function remained normal, and valve appearances were unremarkable. Heparin was reversed with protamine, and decannulation was achieved without complications. The pericardium was closed, and surgical drains were inserted into the pericardial, retrosternal, and right pleural spaces. The sternum was secured with wires, and the skin and soft tissue layers were sutured in the usual manner. The patient maintained sinus rhythm during the entire postoperative period and uneventfully.

The patient recovered well postoperatively, with one episode of paroxysmal atrial fibrillation (PAF) before discharge. He returned home after 1 week and later reported a brief, self-resolving tachycardia. Discharge medications included aspirin 100 mg daily, apixaban 5 mg twice daily (CHADVASC score 3), metoprolol 50 mg twice daily, amiodarone 200 mg daily, rosuvastatin 40 mg at night, and pantoprazole 40 mg daily. At the 5-week follow-up, he was clinically stable (HR 60 bpm, BP 110/70 mmHg), with a normal cardiac exam. Echocardiography showed preserved LV size and systolic function, normal RV size, and mild RV systolic dysfunction. No residual shunt was seen. Blood tests ruled out significant anemia. The cardiologist ceased aspirin and amiodarone. The patient had no further symptoms and was scheduled for review in 6 months.

6 | Discussion

This case demonstrates several noteworthy features of coronary artery fistulae (CAFs) in adults. It highlights that even small or

"silent" CAFs may become clinically relevant over time. The natural history of untreated CAFs shows that while most patients remain asymptomatic during early decades, symptoms often emerge in later life due to progressive enlargement or reduced cardiac reserve [1, 4]. In this patient, a long-standing fistulous connection eventually led to presyncope from coronary steal, in keeping with these findings.

Coronary steal syndrome occurs when blood is preferentially diverted from the myocardium into a low-resistance circuit, reducing perfusion pressure and potentially causing ischaemia or hypotension [2]. Although most cases remain subclinical, documented instances of arrhythmia, infarction, or even cardiac arrest from CAF-related coronary steal have been reported in the absence of obstructive coronary artery disease [12]. It was fortunate that in our patient, symptoms were limited to presyncope, allowing for preemptive surgical correction before irreversible damage occurred. This case also highlights the diagnostic and management challenges associated with CAFs, particularly those detected incidentally. Despite increasing use of imaging modalities, no universal criteria currently exist to guide intervention in asymptomatic cases. Many patients with incidentally found CAFs are managed conservatively, especially in the absence of clinical signs or haemodynamic compromise. However, as demonstrated here, asymptomatic status does not guarantee benignity. Even "silent" fistulae can develop into functionally significant anomalies, especially when patients present with new or unexplained symptoms.

As shown in Table 2, the decision to intervene in CAF depends on a combination of clinical, anatomical, and functional factors. Intervention is typically considered in the presence of symptoms such as chest pain, dyspnoea, or arrhythmia [6, 8], or when imaging suggests a significant left-to-right shunt, potential for ischaemia, or other complications such as rupture, thrombosis, or heart failure [7, 9]. Large or high-flow fistulae, even if asymptomatic, may merit prophylactic closure to reduce future risk [12].

There is a school of thought supporting a watchful waiting approach for small, asymptomatic fistulae due to their often stable nature [2]. Yet long-term complications such as endocarditis, heart failure, and arrhythmias have been reported even in conservatively managed cases [1]. A literature review [12] advised intervention for symptomatic patients and prophylactic closure for large or high-flow CAFs, irrespective of symptoms. Our patient's symptomatic presentation and angiographic findings left little ambiguity—the fistulae warranted surgical correction.

TABLE 2 | Treatment Options for Coronary Artery Fistula.

Treatment option	Description
Percutaneous embolisation [5, 13]	• Minimally invasive; • Uses coils or liquid agents to occlude fistula
Surgical ligation [6, 8]	• Preferred for complex fistulas; • Involves ligation and bypass if needed
Medical management [14]	• For asymptomatic patients; • Includes anti-anginal medications

Treatment options include medical management, transcatheter closure, and surgical ligation. Transcatheter closure is increasingly favored for anatomically suitable cases due to its minimally invasive nature and high success rate [2, 5, 15]. However, complications such as device migration, embolisation, or inadvertent occlusion of native vessels may occur [15]. Surgical intervention remains the preferred option for complex, tortuous, multiple, or diffuse fistulae, especially when arising from multiple coronary territories or when catheter access is not feasible [6, 8, 15, 16].

Medical management is typically reserved for patients with small CAFs who are asymptomatic and show no significant hemodynamic compromise. These patients may be monitored with serial imaging. For those who are symptomatic but unfit for surgery, antianginal medications can be used to manage symptoms and improve quality of life [14].

In this patient, dual fistulae with diffuse origins from the LAD made surgery the more reliable approach. While surgical closure is generally safe, perioperative complications such as thrombosis of the donor vessel must be considered. One series reported myocardial infarction in 11% of cases due to thrombosis in the dilated donor artery following ligation [2]. Antiplatelet therapy, such as aspirin, is often recommended postoperatively to reduce this risk. In our case, the patient was started on aspirin, and follow-up showed no further symptoms and no residual shunting. This has fortunately led to excellent surgical outcomes for CAF, with low perioperative mortality and favorable long-term prognosis, particularly when secondary cardiac damage is absent [16, 17]. Recent case reports also highlight that uncommon anatomical variants, such as a left internal mammary artery–to–pulmonary artery fistula, can lead to coronary steal syndrome, underscoring the importance of high clinical suspicion even in rare presentations [13].

Importantly, this case illustrates the value of persistence and second opinions. Despite earlier reassurance, the patient's decision to seek further evaluation led to a correct diagnosis and appropriate treatment. It can be difficult to revise or challenge a prior specialist's assessment, but evolving symptoms should always prompt re-evaluation. The original conservative approach was reasonable at the time, but the emergence of presyncope tipped the balance toward intervention. The multidisciplinary input and shared decision-making approach ensured patient-centered care and a favorable outcome.

This case reinforces the need for vigilance in managing CAFs. Even long-considered "benign" anomalies may require active intervention if the clinical picture changes. Delayed diagnosis of coronary steal from a CAF could lead to serious consequences. Clinicians should maintain a high index of suspicion, communicate clearly across teams, and remain open to re-examining prior findings in the light of new evidence.

7 | Conclusion

This case highlights key considerations in the management of coronary artery fistulae (CAFs) in adults. It challenges the belief

that small or "silent" CAFs are always benign, demonstrating that asymptomatic lesions can evolve into haemodynamically significant shunts over time. It also reinforces the importance of considering CAF in patients with syncope or ischaemic symptoms, even in the absence of classic risk factors. The patient's progression from stable anatomy to presyncope highlights the value of personalized, multidisciplinary management and the need to revisit prior diagnoses when clinical circumstances change. This case serves as a reminder that ongoing vigilance and a readiness to act on new symptoms are critical to safe and timely care in rare but potentially serious cardiac anomalies.

Author Contributions

Enoch Chi Ngai Lim: conceptualization, data curation, formal analysis, investigation, methodology, project administration, software, validation, visualization, writing – original draft, writing – review and editing. **Chi Eung Danforn Lim:** conceptualization, formal analysis, investigation, resources, supervision, validation, visualization, writing – review and editing.

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Ethics Statement

Patient anonymity is maintained and consent was obtained for publication from the patient.

Consent

Written consent was taken from the patient for publication of this case and accompanying imaging findings.

Conflicts of Interest

The authors declare no conflicts of interest.

Data Availability Statement

Data available on request due to privacy/ethical restrictions.

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