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Interpretation of coronary steal syndrome and haemodynamic changes after surgical closure of coronary fistula using Doppler wire and computational fluid dynamics analysis: a case report

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Background	Coronary arteriovenous fistulas (CAFs) are rare but can cause myocardial ischaemia and other complications. However, the haemodynamic and physiologic characteristics of significant CAFs requiring treatment are poorly described. We report a case of CAF causing coronary steal syndrome in which haemodynamic changes were assessed before and after surgical closure using a Doppler wire and computational fluid dynamics (CFD) technique.
Case summary	A 51-year-old woman presented with exertional chest pain for 3 years. Progressive dyspnoea occurred with exertion. Treadmill and cardiopulmonary exercise tests showed suspicious myocardial ischaemia. Coronary angiography and contrast-enhanced coronary computed tomography angiography (CCTA) revealed a coronary fistula arising from the distal left main coronary artery that drained into the pulmonary artery trunk. We observed a persistent coronary steal phenomenon at baseline and during hyperaemia and a systolic dominant flow rate pattern inside the CAF by Doppler wire-based flow rate measurement. According to CFD analysis based on CCTA, low wall shear stress and a high focal oscillatory shear index were observed at the ostial sites of aneurysmal sacs in the CAF. After successful surgical closure of the CAF, the vessel sizes and flow rate distributions of the coronary arteries increased.
Discussion	Doppler wire-based flow rate distribution measurements and CFD analysis may facilitate the identification of significant coronary steal syndrome requiring closure and the evaluation of future risks of life-threatening complications such as thrombosis and rupture.
Keywords	Coronary steal syndrome • Coronary fistula • Doppler wire • Computational fluid dynamics • Case report

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Learning points

- For the coronary artery fistula (CAF) treatment decision, it is essential to understand 'coronary steal syndrome' and to identify CAF, which is of haemodynamic importance.
- Using Doppler wire-derived flow rate assessment and computed tomography-based computational fluid dynamics technology, we can quantitatively evaluate the effects of CAF on coronary artery circulation.
- After surgical treatment of CAF accompanying a large amount of flow steal, haemodynamic investigations showed increased flow through the coronary arteries and an improvement in cardiopulmonary exercise testing.

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Introduction

Coronary artery fistulas (CAFs) are rarely reported congenital malformations. The incidence of CAFs detected by diagnostic coronary angiography (CAG) is less than 0.2%.[1] Most CAFs originate from the coronary artery, approximately 90% of which drain into the right side of the heart, called arteriovenous fistulas.[2] Coronary artery fistulas can cause myocardial ischaemia, pulmonary hypertension, heart failure, spontaneous intra-pericardial rupture, bacterial endocarditis, and thromboembolism.[3,4] Myocardial ischaemia caused by CAFs is called 'coronary steal syndrome', occurrings when an imbalance exists between the supply and demand of coronary blood flow due to the stealing phenomenon.[4–6] According to current guidelines, closure of CAFs by surgery or intervention is recommended for large CAFs or moderate-size CAFs causing symptoms and clinical complications.[7] However, there are no detailed criteria for distinguishing moderate or large sizes. In addition, various factors such as classifications based on the drainage site, number of fistulous connections, tortuosity, and aneurysmal sacs affect treatment decisions. Therefore, customized treatment approaches are required for asymptomatic patients. We introduce a CAF case treated with surgical closure. In this case, the clinical treatment decisions were made based on precise physiological analysis results. The stealing flow amount through the CAF, indicating myocardial ischaemia was measured by a Doppler-wire, and the haemodynamic characteristics related to the risk of vascular complications were investigated with computational fluid dynamics (CFD).

Timeline

Case presentation

A 51-year-old woman presented with progressive exertional dyspnoea. She had intermittent exertional chest pain for 3 years but was never evaluated. She had a medical history of hyperthyroidism and an old lacunar infarction in the right cerebellum. The initial lab data showed normal N-terminal prohormone of brain natriuretic peptide (99.3 pg/mL) and cardiac enzymes (TnT/CK-MB <3.00 pg/mL/ 0.61 ng/mL). On pre-procedure echocardiography, the left ventricular ejection fraction was 64% and there were no residual wall motion abnormalities. In a treadmill test, 1.4-mm horizontal ST-segment depression in leads II, III, aVF, V5, and V6 developed at peak exercise capacity. On cardiopulmonary exercise (CPEX) testing, decreased exercise function was observed (VO2 65%, VO2/kg 18.67, VE/VCO2 (minute ventilation/carbon dioxide production) 40, 5.3 METs (metabolic equivalents)). CCTA showed no significant coronary stenosis. However, a CAF with tortuous vessel and several markedly dilated aneurysmal sacs was observed at the pre-pulmonic space, and the largest aneurysm was measured to be approximately 10.5 mm × 6.0 mm (*Figure 1A*). The drainage diameter was 2.5–3.1 mm. The differential diagnosis of exertional dyspnoea and chest pain included coronary steal syndrome as a major concern, while variant angina and microvascular disease were less concerning.

To assess coronary haemodynamic changes caused by the CAF, CAG and physiologic assessments were performed. On CAG, the fistula originated from the distal left main (LM) quadrifurcation and drained into the main pulmonary artery trunk (*Videos 1* and *2*). Vasospasm was not induced by an ergonovine provocation test. Resting and hyperaemic average peak velocities (APVs) and coronary

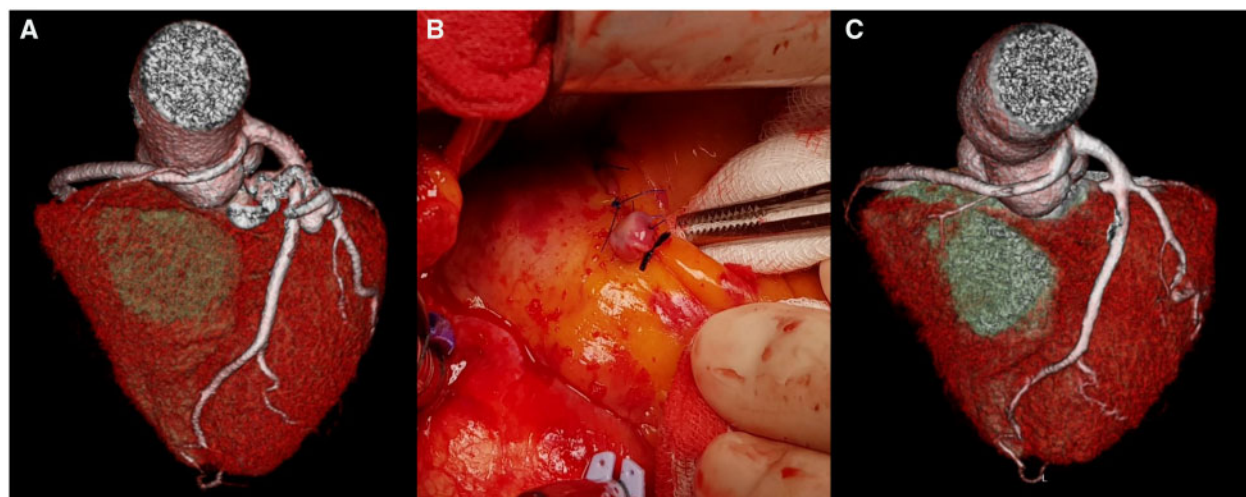| Time	Events
September 2017	A 49-year-old woman had intermittent chest pain since 2017 but was never evaluated.
December 2017	T-wave inversion on the precordial leads of electrocardiography was observed. (incidental finding at the neurology department without further evaluation)
February 2019	The patient came to the cardiovascular centre of the Incheon Saint Mary's Hospital of the Republic of Korea, presenting with persistent chest pain and progressive dyspnoea on exertion. She was diagnosed with a coronary artery fistula (CAF) arising from the distal left main coronary artery and draining into the pulmonary artery trunk using treadmill and cardiopulmonary exercise tests, coronary angiography (CAG), and contrast-enhanced cardiac computed tomography angiography (CCTA). Doppler wire-based flow rate measurements and computational fluid dynamics analysis were performed.
March 2019	Multiple ligations of the CAF were performed with 5-0 Prolene sutures in the on-pump beating state. Follow-up CCTA, CAG, and Doppler wire-based flow rate measurements were performed.
September 2020	Follow-up cardiopulmonary exercise test results were improved.

Figure 1 Images from computed tomography and the operative field. (*A*) Pre-operative 3D computed tomography image showing a coronary fistula with tortuous vessel and several aneurysmal sacs. (*B*) Multiple ligations of the coronary arteriovenous fistula. (*C*) Successful elimination of the coronary arteriovenous fistula on 3D computed tomography.

Video 1 Preoperative coronary angiography video showing a coronary fistula arising from the distal left main coronary artery.
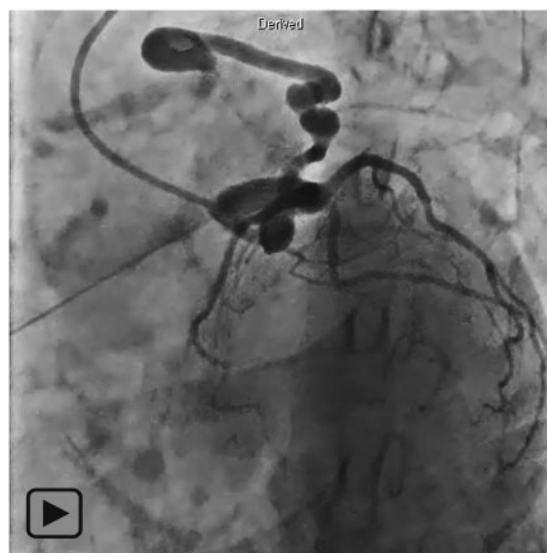

Video 2 Preoperative coronary angiography video showing the distal portion of the coronary fistula draining into the main pulmonary artery trunk.

flow reserve were measured in the coronary arteries [left anterior descending artery (LAD), left circumflex artery (LCX), and ramus intermedius artery (RI)] and CAF using a Doppler wire and a ComboMap System (Philips Volcano, San Diego, CA, USA) (*Table 1*). Based on the radius measured from CCTA and the Doppler-derived APV, the mean aortic absolute flow rate (Qa), mean distal flow rate (Qd), and flow division ratio (Qd/Qa) of each vessel at the LM quadrifurcation area were calculated. The aortic flow rate (Qa) was measured at the entrance of the left main trunk with a catheter. We observed a large amount of blood flow drained by the CAF and a relatively small amount of blood reaching the LAD (*Figure 2*). The Qd/Qa ranking of each vessel showed the same trend both at baseline and during hyperaemia. In addition, we determined the time-averaged wall shear stress (WSS) and oscillatory shear index (OSI) of the fistula by CFD (*Table 1*). Low WSS and focal high-OSI lesions were observed in aneurysmal sacs (*Figure 3A, B*). Due to the size of the CAF, which was larger than 10 mm, a large amount of persistent

Table 1   Equations providing a quantitative estimation of haemodynamics in vessels

Haemodynamic parameter	Definition	Interpretation
Average peak velocity (APV)	$\dfrac{\sum_{n=R_1}^{R_x-1} IPV(n)}{R_x - R_1}$	The APV is the instantaneous peak velocity (IPV) in centimetres per second averaged over the number of beats specified in the APV period of the flow setup screen (between 1 and 5).
Doppler-derived time-averaged flow rate (Q_D)	$\dfrac{\pi D^2}{4}(0.5 \times APV)$	The mean velocity was estimated to be $0.5 \times APV$ by assuming a time-averaged parabolic velocity profile across the vessel. APV is the time-averaged spectral peak velocity. D denotes vessel diameter measured by CCTA.
Time-averaged wall shear stress (WSS)	$\dfrac{1}{T}\int_0^T \tau_w dt$	Time-averaged WSS was calculated by integrating WSS magnitude over the cardiac cycle, in which WSS τ_w is endothelial friction stress by blood flow.
Oscillatory shear index (OSI)	$\dfrac{1}{2}\left[1 - \dfrac{\int_0^T \tau_w dt}{\int_0^T \tau_w dt}\right]$	The oscillatory shear index is a dimensionless metric of changes in the WSS direction.
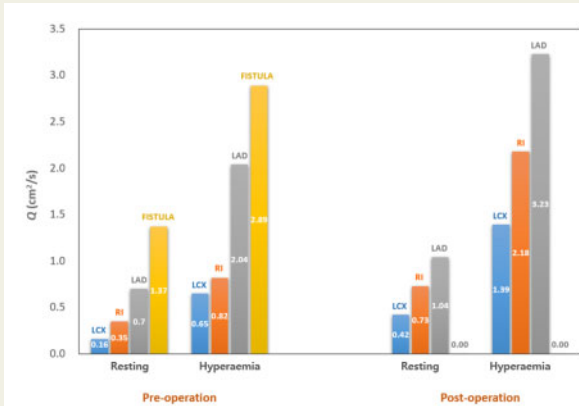
Figure 2 Comparison of coronary anatomy and haemodynamic profiles before and after the operation. We depict the absolute flow rate (Qd) between each vessel and each condition of measurement.

flow steal, extremely high focal OSIs at aneurysmal sacs on CFD analysis, and suspicious ischaemia during the treadmill and CPEX tests, we decided to perform surgical treatment. Computational fluid dynamics analysis was performed using Fluent software (ANSYS, Canonsburg, PA, USA). Multiple ligations of the CAF (*Figure 1B*) were successfully performed. On post-operative CCTA, no remnant fistula or aneurysmal sacs were observed (*Figure 1C*).

Two weeks after the operation, angina assessed by the Seattle angina questionnaire was improved. A treadmill test showed no exercise-induced abnormalities. To assess changes in coronary haemodynamics, CAG and physiologic investigations were performed. The sizes of the coronary arteries (LAD, RI, and LCX) were increased compared to the pre-operative sizes. This finding suggested that the coronary arteries had smaller vessels during the presence of the CAF. We describe the potential mechanism of this condition associated with microvascular resistance and flow steal in Supplementary material online, *Figure S1*. Although the resting and

hyperaemic absolute flowrates in the LM were decreased, the absolute flowrates through the LAD, RI, and LCX were increased, compared with the pre-operative rates (*Figure 2*). The resting and hyperaemic Qd/Qa values reflected an increased flow rate distribution through the LAD, RI, and LCX after surgical treatment. After a year and a half, follow-up CPEX testing showed an enhanced result without remaining symptoms. (VO2 58–92%, METS 4.1–8.5, VE/VCO2 37.7–32.7).

Discussion

Symptomatic CAFs need to be closed by intervention or surgical ligation.[7] However, the approach to asymptomatic moderate to large CAFs remains controversial.[8] Whether CAFs are haemodynamically significant is the key point for management. Nonetheless, the objective amount or cut-off value of stolen coronary blood flow requiring elimination remains poorly investigated. In our case, 'persistent coronary steal' was observed at baseline (Supplementary material online, Figure S1). The proportion of the absolute flow rate in the CAF (53.1%) was higher than that in the LAD (27.1%) (*Figure 2*). After surgical closure of the CAF, the proportion of the LAD flow rate increased to 47.5%. The increase was greater than 20%, indicating the presence of a significant persistent coronary steal that likely induced ischaemia at baseline before surgery.[6,9] In the presence of a CAF, the resistance of the CAF is less than that of the coronary arteries because of the absence of microvascular resistance. Therefore, a shunt to low-pressure structures can dilate the fistula, lower the pressure of the coronary arteries and decrease the size of the LAD. The Qd/Qa rankings of the CAF and LAD did not change between the baseline and hyperaemic states, reflecting residual myocardial ischaemia during hyperaemia. These findings corresponded to the CPEX and treadmill test results indicative of ischaemia.

Normal coronary filling occurs mostly during the diastolic period with a backward suction wave due to ventricular relaxation and relief of myocardial compression.[9] However, in the presence of an arteriovenous CAF, we observed systolic dominant flow with the highest instantaneous peak velocity during the isovolumetric contraction
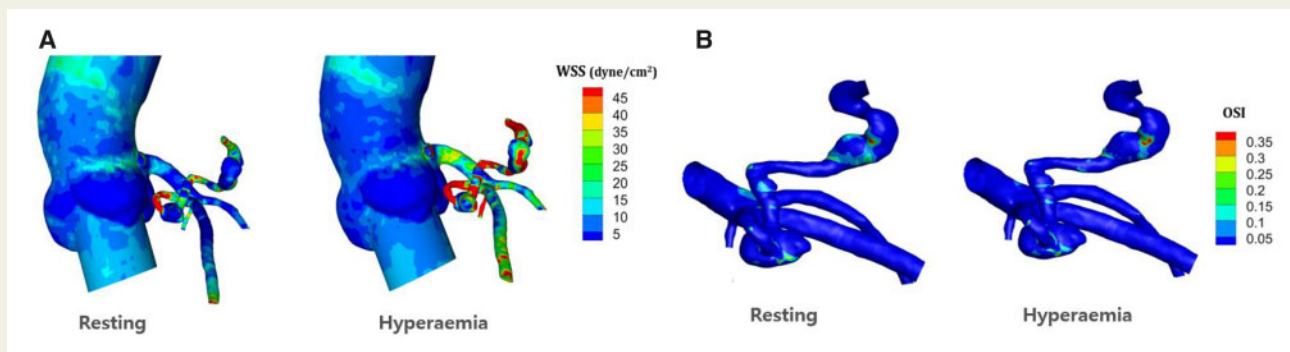
Figure 3 Wall shear stress and oscillatory shear index maps by computational fluid dynamics of the coronary fistula. (*A*) High wall shear stress was observed on the stalk of the fistula where flow was previously accelerated. Low wall shear stress was observed at the site of aneurysmal changes. (*B*) A focal high-oscillatory shear index pattern was observed at the aneurysmal change area in the coronary fistula.
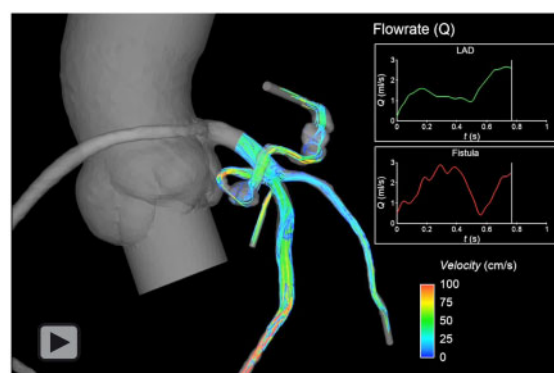
Video 3 Blood flow simulation movie of the left coronary artery in the presence of a coronary fistula artery applying a Doppler wire assisted calculated flow distribution pattern and 3D segmented vessel geometry.

phase (Supplementary material online, *Figure S2*). We simulated the flow distribution in this case during the systolic and diastolic phases using CFD with 3D segmented imaging on CCTA (*Video 3*). We observed a high WSS at the stalk of the CAF, where the flow was accelerated, and low WSS at the sites of aneurysmal changes (*Figure 3A*). High focal regional OSIs were observed in aneurysmal sacs (*Figure 3B*). With these abnormalities of wall shear parameters and the systolic flow dominance of the CAF, we might be able to explain the mechanism of aneurysmal changes in CAFs and the risk of vascular complications, such as intimal ulceration, atherosclerotic deposition, mural thrombosis, and rupture.

Conclusion

We conducted a physiologic evaluation to determine the mechanism of flow rate distribution changes in the presence of a CAF and after the closure of a CAF. Persistent coronary steal was observed at

baseline and during hyperaemia as a result of the stealing pressure because of the low flow resistance of the CAF. The results from Doppler wire measurements and CFD calculations played a decisive role in identifying a haemodynamically significant CAF and determining how to treat it.

Lead author biography

Kwan Yong Lee studied medicine at the Catholic University of Korea. He graduated in February 2009. He completed 4 years of residency and 2 years of fellowship training at Seoul St. Mary's Hospital, Seoul, Korea. Dr. Lee worked as a clinical assistant professor at Incheon St. Mary's Hospital from 2017 to 2020. Since 2021, he has been working as a clinical assistant professor at Seoul St. Mary's Hospital, Seoul, Korea. His main interests are coronary intervention, imaging, and physiology.

Supplementary material

Supplementary material is available at *European Heart Journal—Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidelines.

Conflict of interest: None declared.

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