Coronary Artery Fistula: Current Status

Ye-Hsu Lu
Department of Internal Medicine, Kaohsiung Municipal Siaogang Hospital

Abstract

A coronary artery fistula is an abnormal connection between one of the coronary arteries and another blood vessel or a heart chamber. Cases are usually asymptomatic and of congenital origin, but large fistulae can induce heart failure, myo-ischemia and arrhythmia. Coronary angiography is the main diagnostic tool, and new tools such as computed tomography angiography may enable comprehensive study. Instead of medical therapy, closing the fistula with surgery or intervention can be considered in complicated cases, but complications and recurrence must be taken into account. All patients are different and unique, and any treatment plan should be considered individually.

Keywords: coronary artery fistula, closing device, complication

Introduction

Coronary artery fistula (CAF) is an uncommon entity. It is largely asymptomatic but can lead to several life-threatening complications; hence, appropriate diagnosis and treatment are necessary. In this review, we focus on the evaluation of CAF and the management options available for CAF.

Pathophysiology and Clinical Presentations

A CAF is an abnormal connection between one of the coronary arteries and another blood vessel or a heart chamber. Because CAFs can be asymptomatic and are detected incidentally, the true incidence is difficult to evaluate. The prevalence has been reported to range from 0.2-2%.[1] Studies using computed tomography (CT) angiography show a higher prevalence than studies based on traditional coronary angiography.[2]

CAFs usually have various and complicated image features, and the clinical symptoms mainly depend on the size, origin and drainage site of the fistulae.[3]

Most patients with small CAFs are asymptomatic and may experience spontaneous closure during long-term follow-up. Therefore, management of CAF depends on the significance of symptoms.[4] Factors that determine the hemodynamic significance of the fistulous connection include the size of the communication, the resistance of the recipient chamber, and the potential for development of myocardial ischemia.[5] CAFs may mimic the physiology of various heart lesions. Fistulae that (1) drain to the systemic veins or right atrium generate a

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Address for correspondence: Ye-Hsu Lu, MD
Department of Internal Medicine, Kaohsiung Municipal Siaogang Hospital; No. 482, Shanming Rd., Siaogang Dist., Kaohsiung City 812, Taiwan
Tel: +886-7-8036783, +886-918867017; E-mail: yehslu@kmu.edu.tw

physiology similar to an atrial septal defect; (2) those that drain to the pulmonary arteries have a physiology similar to a patent ductus arteriosus; (3) those draining to the left atrium do not cause a left to right shunt but do cause a volume load similar to mitral regurgitation; and (4) those draining to the left ventricle produce a physiology similar to that of aortic insufficiency.[6]

Etiology and Classification

There are three major causes of CAF, whereby the most common is congenital origin. Acquired CAF may come from trauma, such as gunshot wounds or stab injuries. Iatrogenic causes are becoming more common as a result of increasing interventional or surgical procedures.[7] Table 1 summarizes the rules for CAF classification.[8] Frequently, the feeding artery of the CAF may drain from a coronary artery or one of its branches after a tortuous and dilated course that ends in one of the cardiac chambers or in a vessel. Figure 1 illustrates the morphology of fistula origins, drainage sites, and numbers of drainage tracts.[9] The size of CAFs is classified by comparing with neighboring non-feeding vessels. If the diameter of a feeding vessel is less than the neighboring non-feeding vessels, the size of the CAF is defined as small. A medium size CAF is within 1-2 times the diameter of neighboring non-feeding vessels, while a large CAF has over twice that size.[10] The drainage vessels may be single or multiple, and may be straight or tortuous. Table 2 shows the frequencies of origins and drainage

Table 1. Rules for CAF classification

Classification	Descriptions
Based on etiology	Congenital Acquired: Iatrogenic, disease-related, trauma-related
Based on origin of feeding artery	From RCA From LCA: LAD, LCX, or other main branches From both RCA and LCA From other anomalous coronary arteries
Based on segment of fistula origin	Sakakibara classification Type A: Originating from proximal native vessel, distal artery is normal Type B: Originating from distal vessel and entire proximal coronary artery is dilated
Based on drainage site	Coronary cameral fistula: involving any cardiac chamber Coronary artery fistula: involving pulmonary artery, coronary sinus, SVC, IVC, bronchial vessels, other extracardiac veins
Based on number of fistulous tracts	Single or multiple
Based on morphology and complexity	Simple: single origin, straight course, and single drainage site Complex: multiple fistulous structure or entangled vessel course
Based on accompanying anomaly	Isolated Fistula accompanied by other structural abnormalities

RCA: right coronary artery, LCA: left coronary artery, LAD: left anterior descending artery, LCX: left circumflex artery, SVC: superior vena cava, IVS: inferior vena cava sites of CAFs.[11] Most frequently, they arise from the right coronary artery (50-60%) and most often drain into the right heart (over 80%). Most CAFs are unilateral, but multiple feeding vessels should be carefully evaluated. CAFs can be classified as proximal or distal, based on the site of fistula origin from the conduit coronary artery (Figure 2).[12] The proximal conduit of a distal type CAF may dilate gradually during long-term follow-up.[4]

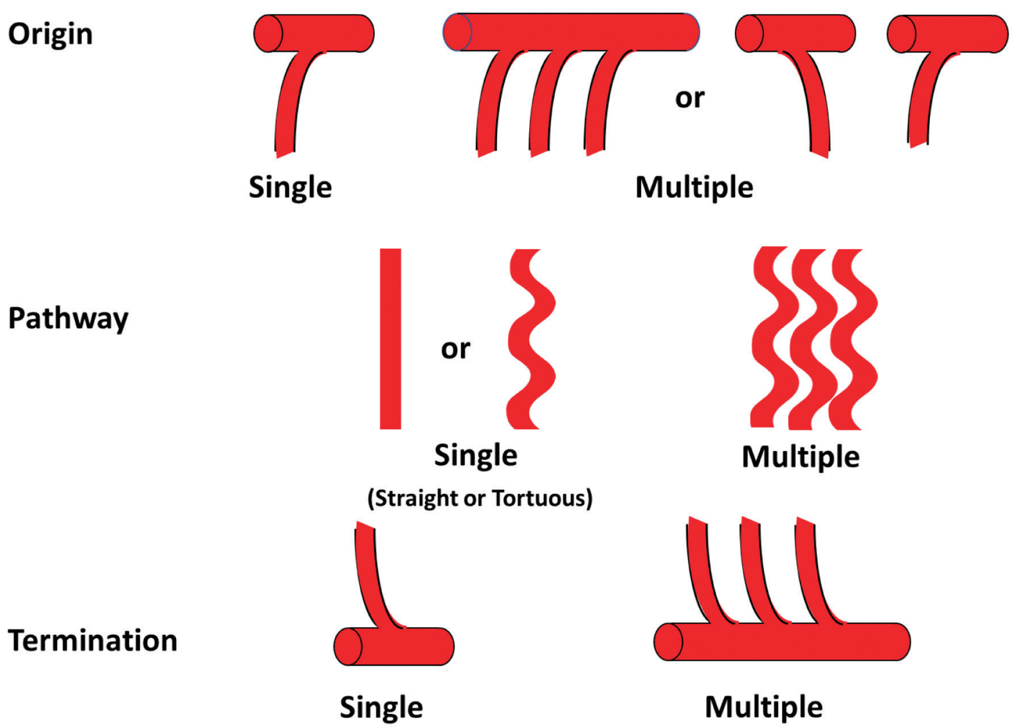

Figure 1. Morphology of origins, pathways and terminations of CAFs. (Adapted from Said S.A. et al.[9])

Table 2. Frequency of origins and drainage targets of CAFs

Origin	Prevalence	Drainage	Prevalence
LAD	25-42	RA	19-26
RCA	50-60	RV	14-40
Both	5	LA	5-6
Diagonal branch	1.9	LV	2-19
Marginal branch	0.7	PA	15-20.2
Circumflex	18.3	CS	7
Single coronary	3	SVC	1

RA: right atrium, RV: right ventricle, LA: left atrium, LV: left ventricle, PA: pulmonary artery, CS: coronary sinus, SVC: superior vena cava.

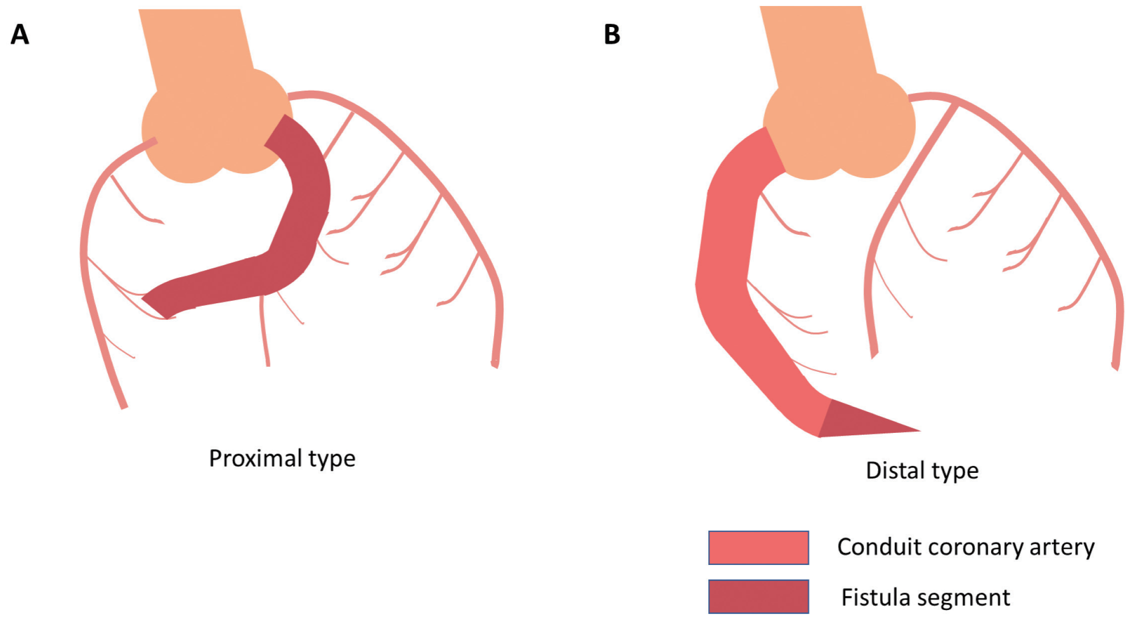

Figure 2. Proximal and distal type of CAF. A: Proximal CAF originates from the proximal epicardial coronary artery, and the long fistula segment is devoid of coronary branches. B: Distal fistula originates from the distal epicardial artery, and the conduit epicardial artery proximal to the last coronary branch is dilated with normal coronary branches. (Adapted from Gowda ST. et al.[12])

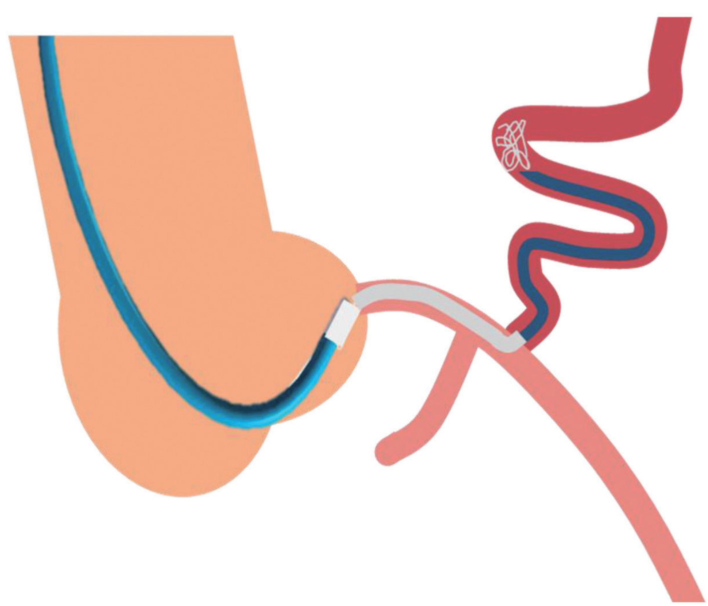

Figure 3. Telescoping technique for coil delivery. A guide catheter (blue) is used to engage the left main coronary artery. The microcatheter (gray) is advanced over the guidewire up to the origin of the left anterior descending artery to the pulmonary artery fistula. With the microcatheter support, a smaller microcatheter wire (dark blue) is advanced over the guidewire into the fistula, followed by coil delivery. (Adapted from Sulemankhil I. et al.[24])

Clinical Manifestations and Evaluations

Children with CAF are mostly asymptomatic (80%), while about 40% of adults share symptoms of effort fatigue, dyspnea (60%), angina and congestive heart failure.[13]

Table 3 shows the frequency of possible symptoms in patients with CAF, including symptoms of heart failure, endocarditis and myo-ischemia.[14] Arrhythmias with or without myo-ischemia are uncommon, but some may be life-threatening.[15] Echocardiography is valuable in evaluating cardiac dysfunction and revealing the origin of a CAF. The diagnosis of coronary fistula can usually be made by 2D echocardiography with color flow mapping.[16] However, using pure 2D echocardiography without Doppler study it may be more difficult to detect small or even multiple CAFs.[17] And even with the use of Doppler echocardiography to detect CAFs, the observations can be confounded by a long list of possible flows from abnormal structures to the pulmonary artery, including ductus arteriosus, coronary artery aneurysm, pulmonary arteriovenous fistula, ruptured sinus of Valsalva aneurysm, aortopulmonary window, prolapse of the right aortic cusp with a supracristal ventricular septal defect, etc., etc.….[18] While coronary angiography has traditionally been used to diagnose CAF, the precise course of CAF anatomy can be difficult to delineate, particularly the drainage sites and tortuous courses. CT angiography may provide additional information about spatial structure of CAFs with high temporal resolution.[19] Table 4 summarizes the items that are suggested for evaluation before and after procedures.[20]

Table 3. Frequency of CAF symptoms presented.[13]

Symptoms	Frequency (cases %)
Dyspnea	Common (60%)
Endocarditis	Good (20%)
Congestive heart failure	Good (19%)
Angina	Moderate (3-7%)
Syncope	Uncommon
Palpitations	Uncommon
Cardiac arrhythmias	Uncommon
Hemopericardium	Rare
Cardiac tamponade	Rare
Sudden cardiac death	Isolated

Treatments

Table 5 summarizes the indications of various treatments for CAFs.[20] The management of CAFs depends on their size and symptomatology. The guidelines recommend that small,

Table 4. Items for evaluation before and after procedures.[20]

Preprocedural evaluation	Postprocedural evaluation
Number and location of fistula origins	Residual leakage or recanalization
Number and location of drainage sites	Aneurysmal change or persistent dilatation of coronary artery and ostium
Courses and size of fistula tract	Thrombus formation
Fistula complexity: Angulations, tortuosity, associated connection with other structures	Coil or device migration
Other associated anomalies	Myocardial ischemia or infarction
	Catheter-related complications
	Coronary artery spasm, dissection or perforation

Table 5. Indications for the various treatment modalities for CAFs.[20]

Treatments	Indications
Medications	Small CAFs Medium-sized asymptomatic CAFs
Trans-catheter closure	Proximal CAFs with single –site drainage Distal narrowing of CAF with feasible device access Non-tortuous vessels and absence of vessels that could be embolized accidentally Serious cardiac co-morbidities
Surgery	Cases requiring concomitant surgical procedures Distal CAFs High-velocity CAFs CAFs with tortuous course or complex connections Those with multiple drainage sites or a "fistula lake" Signs of volume overload Proximal CAF with large aneurysms

asymptomatic CAFs be treated conservatively and monitored with echocardiograms every 3-5 years in case of enlargement, degeneration or symptomatic progression. Larger fistulae have a higher risk of complications and should be closed either percutaneously or surgically, even if asymptomatic. Small to moderate fistulae should only be closed if symptomatic or in the presence of complications such as unexplained structural changes or dysfunction, high shunt flow or aneurysmal degeneration.[21,22] Al-Hijji M. et al. have reviewed the benefits and risks of non-invasive and invasive treatments for CAF.[23] Surgical procedures are especially suitable for significant symptomatic shunt flow, or if concomitant structural abnormality should be treated at the same time. Trans-catheter closure of CAFs is noted for its easier approach and implanting of the closing devices. Newly developed delivery systems are highly recommended for better support; for example, the "telescoping technique" using microcatheters can be applied for coil implantation.[24] Table 6 compares the CAF sizes with the available devices for CAF closure, recommended delivery methods, delivery approaches, and required levels of clinical experience.[25]

Unfortunately, recanalization of CAFs is observed after both surgical and trans-catheter closures and slow flow, thrombosis and myocardial infarction have been reported.[10,14,23,26] Oral anticoagulation is indicated for the prevention of myocardial infarction in high-risk patients.[23]

Regular follow-up after disease confirmation and treatment is necessary (Table 4).[20] Comprehensive evaluation by a heart team that includes a pediatric or general cardiologist familiar with the patient and an interventional cardiologist and cardiothoracic surgeon with expertise in fistula closure should take into account the patient's preferences and comorbidities.

Conclusions

CAF is a relatively rare condition with heterogenous presentations. Advances in cardiac imaging can improve the accuracy of diagnosis and development of the treatment plan. Thorough discussion between the patient and the members of the heart team are beneficial for decision making.

Table 6. Comparison of devices used in percutaneous coronary fistula closure.[25]

	Amplazer vascular plug	Amplazer duct occluder	Detachable balloon	Coils	Cover stent	Chemicals
Fistula size	M-L	M-L	M-L	S-M	S-M	S-M
Retraction before deployment	Y	Y	Y	Y/N	N	N
Required clinical experience	High	High	Low	High	Moderate	Low
Delivery approach	Antegrade	Antegrade Retrograde	Retrograde	Antegrade Retrograde	Retrograde	Retrograde

S: small size, M: medium size, L: large size, Y: yes, N: no

Regular follow-up after treatment is important for early detection of possible complications.

References

1. Matsumoto Y, Kawano H, Iwasaki K, et al. Histopathology of giant coronary artery aneurysm associated with coronary artery fistula. *Int Heart J* 2018;59:431-434.
2. Lim JJ, Jung JI, Lee BY, et al. Prevalence and types of coronary artery fistulas detected with coronary CT angiography. *Am J Roentgenol* 2014;203:W237-243.
3. Li N, Zhao P, Wu D, et al. Coronary artery fistulas detected with coronary CT angiography: a pictorial review of 73 cases. *Br J Radiol* 2020;93:20190523.
4. Lo MH, Lin IC, Hsieh KS, et al. Mid- to long-term follow-up of pediatric patients with coronary artery fistula. *J Formos Med Assoc* 2016;115:571-576.
5. Sagar D, Hernandez A, Heimowitz T. Coronary artery-left ventricle fistula: a case report of a rare connection error! *Cureus* 2015;7:e266.
6. Latson LA. Coronary artery fistulas: how to manage them. *Catheter Cardiovasc Interv* 2007;70:110-116.
7. Sunkara A, Chebrolu LH, Chang SM, et al. Coronary artery fistula. *Methodist Debakey Cardiovasc J* 2017; 13:78-80.
8. Reddy G, Davies JE, Holmes DR, et al. Coronary artery fistulae. *Circ Cardiovasc Interv* 2015;8: e003062.
9. Said SA, van der Werf T. Dutch survey of coronary artery fistulas in adults: congenital solitary fistulas. *Int J Cardiol* 2006;106:323-332.
10. El-Sabawi B, Al-Hijji MA, Eleid MF, et al. Transcatheter closure of coronary artery fistula: a 21-year experience. *Catheter Cardiovasc Interv* 2020;96:311-319.
11. Challoumas D, Pericleous A, Dimitrakaki IA, et al. Coronary arteriovenous fistulae: a review. *Int J Angiol* 2014;23:1-10.
12. Gowda ST, Latson L, Sivakumar K, et al. Anatomical classification and posttreatment remodeling characteristics to guide management and follow-up of neonates and infants with coronary artery fistula: a multicenter study from the coronary artery fistula registry. *Circ Cardiovasc Interv* 2021;14:e009750.
13. Liberthson RR, Sagar K, Berkoben JP, et al. Congenital coronary arteriovenous fistula. Report of 13 patients, review of the literature and delineation of management. Circulation 1979;59:849-854.
14. Buccheri D, Chirco PR, Geraci S, et al. Coronary artery fistulae: anatomy, diagnosis and management strategies. *Heart Lung Circ* 2018;27:940-951.
15. Ghumman GM, Khan A, Shafqat M, et al. A coronary cameral fistula associated with incessant ventricular arrhythmias. *Cureus* 2023;15:e35847.
16. Barbosa MM, Katina T, Oliveira HG, et al. Doppler echocardiographic features of coronary artery fistula: report of 8 cases. *J Am Soc Echocardiogr* 1999;12:149-154.
17. Xie M, Li L, Cheng TO, et al. Coronary artery fistula: comparison of diagnostic accuracy by echocardiography versus coronary arteriography and surgery in 63 patients studied between 2002 and 2012 in a single medical center in China. *Int J Cardiol* 2014;176:470-477.
18. Gowda RM, Vasavada BC, Khan IA. Coronary artery fistulas:clinical and therapeutic considerations. *Int J*

Cardiol 2006;107:7-10.
19. Ali A, Colledge J, Sri I, Missouris C. CT: the imaging of choice in the diagnosis of coronary artery fistulae. *BJR Case Rep* 2016;2:20150492.
20. Yun G, Nam TH, Chun EJ. Coronary artery fistulas: pathophysiology, imaging findings, and management. *Radiographics* 2018;38:688-703.
21. Warnes CA, Williams RG, Bashore TM, et al. ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association task force on practice guidelines (writing committee to develop guidelines on the Management of Adults with congenital heart disease). Developed in collaboration with the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons. *J Am Coll Cardiol* 2008;52: e143-e263.
22. Baumgartner H, De Backer J, Babu-Narayan SV, et al. 2020 ESC Guidelines for the management of adult congenital heart disease. *Eur Heart J* 2021;42:563-645.
23. Al-Hijji M, El Sabbagh A, El Hajj S, et al. Coronary artery fistulas: Indications, techniques, outcomes, and complications of transcatheter fistula closure. *JACC Cardiovasc Interv* 2021;14:1393-1406.
24. Sulemankhil I, Mohamed A H, Gilani S A. Coronary-pulmonary artery fistula repair with coil embolization: a single center experience. *Cureus* 2022;14:e28407.
25. Oto, M.A., Yorgun, H., Aytemir, K. Percutaneous approaches to closure of coronary artery fistulas. *Interventional Cardiology* 2011;3:79-90.
26. Said SM, Burkhart HM, Schaff HV, et al. Late outcome of repair of congenital coronary artery fistulas -- a word of caution. *J Thorac Cardiovasc Surg* 2013; 145:455-460.