JACC: CASE REPORTS VOL. 4, NO. 9, 2022

CASE REPORT

ADVANCED

CLINICAL CASE

Transcatheter Closure of Complex Left Circumflex to Coronary Sinus Fistula

Nanda K. Pullela, MBBS,[a] Deepak Acharya, MD, MSPH,[b] Madhan Shanmugasundaram, MD,[b] Tushar Acharya, MD, MPH,[b] Muhammad Ajmal, MBBS,[b] Huu T. Truong, MD,[c] Ranjith Shetty, MD,[d] Kapil D. Lotun, MD[d]

ABSTRACT

Coronary artery fistulae connecting the left circumflex to the coronary sinus are rare. Surgical closure of coronary sinus connections is technically challenging because of the location, especially in high-risk surgical patients. We used multimodality imaging to delineate the drainage site and successfully closed a left circumflex to coronary sinus fistula using a transcatheter closure technique. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2022;4:559-563)

Coronary artery fistula (CAF) is a direct communication of a coronary artery with a cardiac chamber, great vessel, or other vascular structure, bypassing the myocardial capillary bed. CAF is rare, affecting 0.002% of the general population, and represents about 0.2% to 0.4% of all cardiac malformations and 14% of all coronary anomalies.[1] The hemodynamic consequences of CAF depend on the different pressures between the coronary artery and the site where the fistula drains.[1] The most frequent draining sites of CAF are the right ventricle (RV), the proximal pulmonary artery (PA), and the right atrium (RA).

LEARNING OBJECTIVES

- To consider coronary artery fistulae as a rare cause of heart failure presentation in elderly.
- To understand the importance of multimodality imaging in preprocedural planning for closure of complex coronary artery fistula.
- To foresee technical challenges during closure of coronary artery fistula using transcatheter closure techniques.

HISTORY OF PRESENTATION

A 72-year-old man presented with progressively worsening dyspnea and bilateral leg swelling for 3 months.

MEDICAL HISTORY

The patient had a history of persistent atrial fibrillation, hyperlipidemia, hypertension, gastroesophageal reflux disease, and chronic obstructive airway disease.

DIFFERENTIAL DIAGNOSIS

Given the patient's worsening dyspnea and leg swelling, congestive heart failure, cor pulmonale, and

From the [a]Department of Cardiology, Banner University Medical Center, Tucson, Arizona, USA; [b]Department of Cardiology, Banner University Medical Center, Tucson, Arizona, USA; [c]Department of Cardiology, Loma Linda University Medical Center, Loma Linda, California, USA; and the [d]Department of Cardiology, Carondelet Heart and Vascular Institute, Tucson, Arizona, USA.
The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

Manuscript received February 23, 2022; accepted March 17, 2022.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2022.03.017

ABBREVIATIONS AND ACRONYMS

CAF = coronary artery fistula
CS = coronary sinus
LCx = left circumflex
PA = pulmonary artery
RA = right atrium
RV = right ventricle
TCC = transcatheter closure

tachycardia-induced cardiomyopathy were considered in the differential diagnosis.

INVESTIGATIONS

His baseline electrocardiogram showed atrial fibrillation with no ST, T changes. A transthoracic echocardiogram showed severe RA and RV enlargement and moderate RV dysfunction. There was tricuspid annular dilatation and leaflet noncoaptation, with severe tricuspid regurgitation. Color Doppler imaging showed increased flow in the coronary sinus (CS) (Video 1). The RV systolic pressure was estimated at 27 mm Hg. The inferior vena cava was dilated and noncollapsible. The left ventricular size was normal, with mildly impaired systolic function and an ejection fraction of 45% to 50% along with dilation of the left atrium.

Right heart catheterization showed a markedly elevated RA pressure of 22 mm Hg, with a prominent V-wave, RV end-diastolic pressure of 18 mm Hg, mean PA pressure of 27 mm Hg and pulmonary capillary wedge pressure of 22 mm Hg. There was a step-up in the oxygen saturation from 60% in the superior vena cava and inferior vena cava, respectively, to 69% in the mid-RA and 72% in the RV and the PA. The calculated pulmonary-to-systemic flow ratio was 1.5.

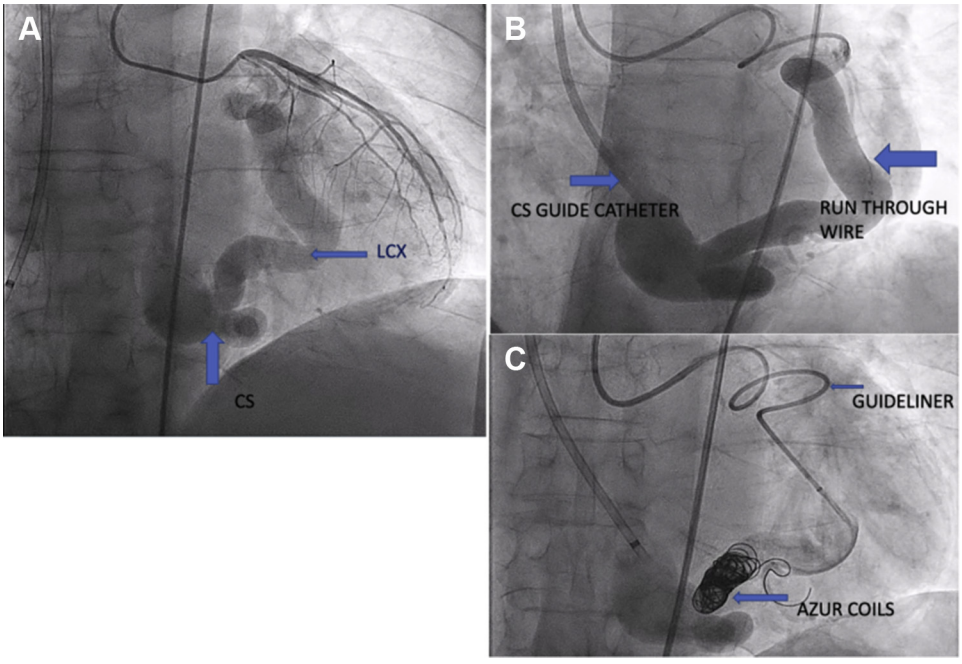
Coronary angiography showed marked dilation of left main artery. The left circumflex coronary artery (LCx) was markedly dilated and tortuous, draining directly into the CS in a manner suggestive of an LCx-to-CS fistula (**Figure 1A,** Video 2). The right coronary artery was a nondominant small vessel.

Computed tomography coronary angiography confirmed the findings of a large distal LCx-to-CS fistula (**Figures 2 and 3**). The LCx measured 1.4 cm in the AV groove, and the fistula measured 1.2 cm in caliber, respectively. Cardiac magnetic resonance demonstrated the tortuous and severely dilated left circumflex artery and dilated CS, moderate RV enlargement, and moderate dysfunction (Videos 3 to 5).

MANAGEMENT

Based on the multimodality imaging, the patient had a large distal CAF that caused hemodynamically significant left-to-right shunting with RA, RV, LA dilatation and right heart failure. He was considered to be at a high surgical risk, given his age and comorbid condition, and thus percutaneous repair was planned.

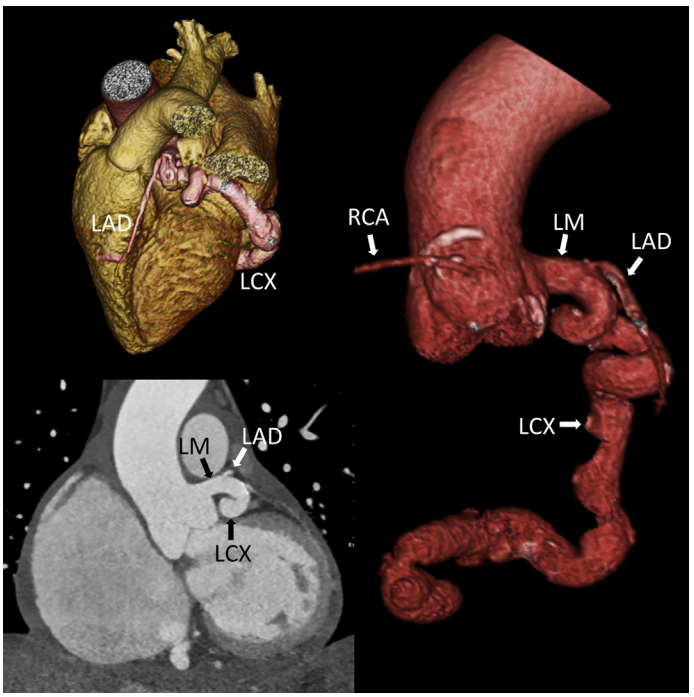
FIGURE 1 Angiographic Images of Fistula Cosure



(A) Coronary angiography in left anterior oblique view demonstrating left circumflex (LCx) to coronary sinus (CS) fistula. **(B)** Fistula is negotiated with coronary wire and CS guide catheter engaged in proximal CS. **(C)** Successful deployment of coils via arterial approach.

The patient was brought into the cardiac catheterization laboratory. Both groins and the right side of the neck were prepared. An 11-F sheath was placed in the right internal jugular vein. A 7-F sheath was placed in the right common femoral artery. A 7-F extra backup 4.0 guide catheter (Medtronic) was used to intubate the left main coronary artery. Heparin intravenous boluses were administered to target activated clotting time >250 seconds. The procedural plan was to deploy a vascular plug from a venous approach at the distal entry of the fistula into the CS and to place coils via the arterial approach at the narrowest portion of the fistula to prevent distal coil embolization. The LCx-to-CS fistula was negotiated with Runthrough (Terumo) and Glidewire (Terumo). For the arterial approach, we telescoped a 7-F Guideliner (Vascular Solutions Inc) and a 4-F Glidecath/Progreat (Terumo) microcatheter distally into the narrowest part of the fistula (**Figure 1B,** Video 6). For the venous approach, we upsized the right internal jugular vein access to a steerable CS guide catheter and engaged the proximal CS (**Figure 1B,** Video 6). Attempts to negotiate the distal fistula with an angled hydrophyilic-coated guidewire on various hydrophilic-coated catheters in multiple projections were unsuccessful because of the complex lobular anatomy with multiple acute bends. As such, we were unable to deliver a vascular plug via the venous approach. From the arterial access via the coronaries, several attempts were made to navigate the wires from the fistula into the CS, which were unsuccessful. Ultimately, we decided to coil embolize the fistula via the arterial approach. We delivered a 0.018-inch Azur CX (Terumo) 16/39-mm, 16/39-mm, 14/34-mm, 10/32-mm, 12/38-mm, 12/38-mm, and 10/32-mm coils into the distal fistula through a microcatheter. The coils were packed nicely into an area of narrowing with stable positioning (**Figure 1C,** Video 7). Care was taken to deploy the coils distal to the last obtuse marginal branch to prevent thrombosis and subsequent myocardial injury. A single loose end of a coil that embolized into the small OM branch was pulled back and repositioned with a 5.0-mm balloon. Repeated angiography of the fistula demonstrated near-contrast stasis with mild residual flow. The patient did not have any procedural complications.

FIGURE 2 Reformatted Computed Tomography Coronary Angiography



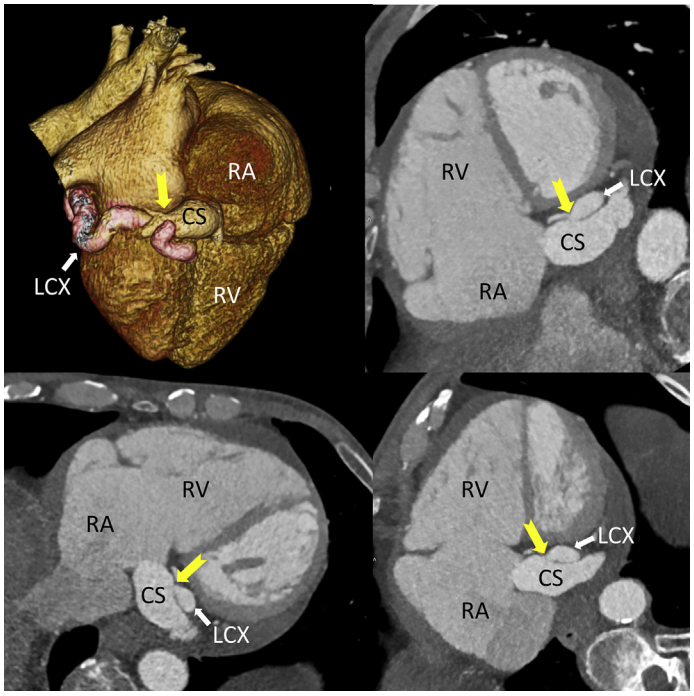
Virtually rendered and coronal computed tomography images show a diffusely aneurysmal left main (LM) and left circumflex (LCx) coronary artery with a relatively normal caliber left anterior descending (LAD) coronary artery. RCA = right coronary artery.

DISCUSSION

CAFs are usually asymptomatic in the majority of infants and children but tend to cause symptoms in individuals >20 years of age. Presenting symptoms are fatigue, dyspnea, angina, and congestive heart failure.[2] Our patient had a large distal LCx-to-CS fistula resulting in predominant right-sided chamber dilatation and right heart failure. Pretricuspid drainage of fistulae results in RA/RV dilatation. Even though the fistula was large, the pulmonary-to-systemic flow ratio was 1.5. This discordance, caused by difficulty in oximetric shunt measurement, has been noted by others.[3] A fistula terminating in the CS is known to be the sole anatomical risk factor for postrepair (surgical or percutaneous) adverse events such as coronary thrombosis, myocardial infarction, and cardiomyopathy.[4] Transcatheter closure (TCC) or surgical closure is a Class I recommendation for large fistulae regardless of symptoms and for small to moderate-size fistulae with evidence of myocardial ischemia, arrhythmia, ventricular dysfunction, ventricular enlargement, or endarteritis.[5] Surgical repair is often preferred when the fistula is large and tortuous, with distal connections to the low-pressure chambers. TCC of a CAF is unsuitable when there is extreme vessel tortuosity, multiple drainage sites, and coronary branches at the site of optimal device positioning.[6] CS connections are difficult to close completely even with open surgical repair because of the presence of multiple connections at times and the location of the connection on the posterior base of the heart.

JACC: CASE REPORTS, VOL. 4, NO. 9, 2022
MAY 4, 2022:559–563

FIGURE 3 Reformatted Cross-Sectional Computed Tomography Coronary Angiography



Virtually rendered and multiplanar reformatted contrast-enhanced coronary computed tomography angiogram images show the aneurysmally dilated left circumflex artery (LCx) draining into a dilated coronary sinus (CS), representing an LCx-to-CS fistula. RA = right atrium; RV = right ventricle.

Our patient presented with symptoms of right heart failure in his eighth decade, which is similar to the late age at presentation described by Valente et al.[4] He presented a technical challenge for TCC because of a dilated, tortuous LCx with complex lobular anatomy of the fistula and a coronary branch supplying normal myocardium close to the planned coil deployment site. Collins et al.[7] described vessel tortuosity and lumen caliber as significant limitations in occlusive device delivery. Attempts at a venous approach to the distal fistula through the CS guide catheter was unsuccessful. Therefore, the fistula was closed with multiple coils delivered through a microcatheter, distal to a small coronary branch. Gowda et al[8] showed that a large distal fistula with persistent proximal conduit vessel dilatation can cause flow stasis and thrombosis after CAF closure and suggested long-term anticoagulation. Our patient was discharged to take apixaban. Large distal fistula and age at closure are important risk predictors for thrombosis risk. Consideration for closure should be based on the severity of symptoms versus the risk of thrombosis after closure.[9] Residual flow would be beneficial with a large distal fistula closure because it would offset the stasis and associated risk of thrombosis in the proximal dilated conduit vessel.[3,8] Our patient had small residual flow after the procedure, which persisted at the 6-month follow-up angiogram. Shah et al[3] proposed a long-term angiographic follow-up after CAF closure.

FOLLOW-UP

After discharge, the patient continued to take his apixaban, which he had been taking for his atrial fibrillation. The patient had a significant improvement in his symptoms 1 month after the procedure. His 6-month follow-up selective angiogram showed mild residual flow through the fistula with no evidence of proximal thrombus extension into the conduit vessel (Video 8).

CONCLUSIONS

The optimal treatment of a large distal CAF is a topic for continued debate. The benefit of closure versus the long-term risk of thrombosis should be weighed before closure is attempted. This case not only posed a technical difficulty at TCC but challenged our decision to close.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

ADDRESS FOR CORRESPONDENCE: Dr Nanda K. Pullela, Sarver Heart Center, 1501 N. Campbell Avenue, Tucson, Arizona 85724, USA. E-mail: nandapk89@gmail.com.

REFERENCES

1. Dario B, Paolo RC, Salvatore G, Giuseppe C, Bernardo C. Coronary artery fistulae: Anatomy, Diagnosis and Management Strategies. *Heart Lung Circ*. 2018;27:940–951.

2. Liberthson RR, Sagar K, Berkoben JP, Weintraub RM, Levine FH. Congenital coronary arteriovenous fistula: report of 13 patients. Review of literature and delineation of management. *Circulation*. 1979;59:849–854.

3. Shah AH, Osten M, Benson L, Alnasser S. Long-term outcomes of percutaneous closure of coronary artery fistulae in the adult: A single-center experience. *Catheter Cardiovasc Interv*. 2020;95:939–948.

4. Valente AM, Lock JE, Gauvreau K, et al. Predictors of long-term adverse outcomes in patients with congenital coronary artery fistulae. *Circ Cardiovasc Interv*. 2010;3:134–139.

5. 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. *J Am Coll Cardiol*. 2008;52(23):e143–263.

6. Armsby LR, Keane JF, Sherwood MC, Forbess JM, Perry SB, Lock JE. Management of coronary artery fistulae. Patient selection and results of transcatheter closure. *J Am Coll Cardiol*. 2002;39(6):1026–1032.

7. Collins N, Mehta R, Benson L, Horlick E. Percutaneous coronary artery fistula closure in adults: technical and procedural aspects. *Catheter Cardiovasc Interv*. 2007;69(6):872–880.

8. Gowda ST, Latson LA, Kutty S, Prieto LR. Intermediate to long-term outcome following congenital coronary artery fistulae closure with focus on thrombus formation. *Am J Cardiol*. 2011;107:302–308.

9. Gowda ST, Forbes TJ, Singh H, et al. remodeling and thrombosis following closure of coronary artery fistula with review of management: Large distal coronary fistula- To close or not to close? *Catheter Cardiovasc Interv*. 2013;82:132–142.

KEY WORDS coronary artery fistula, transcatheter closure

APPENDIX For supplemental videos, please see the online version of this article.