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CASE REPORT

ADVANCED

CLINICAL CASE

Closing a Right Coronary Artery Fistula Draining Into the Coronary Sinus Using a Covered Stent in the Coronary Sinus

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ABSTRACT

This report describes the case of a symptomatic patient with a right coronary artery fistula draining into the coronary sinus who underwent transcatheter closure, which was deployed in the drainage site to seal off the exit of the fistula. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2021;3:1589–1593) © 2021 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 68-year-old previously asymptomatic female developed palpitations based on atrial fibrillation (AF), which were treated at the outpatient clinic with direct oral anticoagulation therapy and beta blockers.

LEARNING OBJECTIVES

- To recognize CAFs as a potentially significant burden resulting in heart failure symptoms after remaining asymptomatic for a long period of time.
- To choose transcatheter closure as a treatment option for symptomatic CAFs, instead of surgical closure if technically feasible with an acceptable risk.
- To use covered stents to close fistulae by deployment in the drainage site, instead of the feeding artery.

A week later her symptoms worsened with dyspnea and orthopnea, for which she was hospitalized. Physical examination showed a blood pressure of 113/71 mm Hg, an irregular pulse and a normal jugular venous pressure. On auscultation, no (continuous) murmur of the heart was heard, and the lungs were normal. The electrocardiogram showed AF with a high ventricular response rate.

MEDICAL HISTORY

In addition to AF, which was of unknown duration and presumably present already for a longer time, the patient had no other relevant medical history.

DIFFERENTIAL DIAGNOSIS

The clinical hypothesis was heart failure due to tachycardiomyopathy caused by long-standing atrial arrhythmia. Differential diagnostic considerations

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ABBREVIATIONS AND ACRONYMS

ACHD = adults with congenital heart disease

AF = atrial fibrillation

CAF = coronary artery fistula

CS = coronary sinus

RCA = right coronary artery

RDP = ramus descendens posterior

RPL = ramus posterolateralis

were primary or secondary valvular heart diseases, coronary artery disease, hypertension, and pulmonary embolism.

INVESTIGATIONS

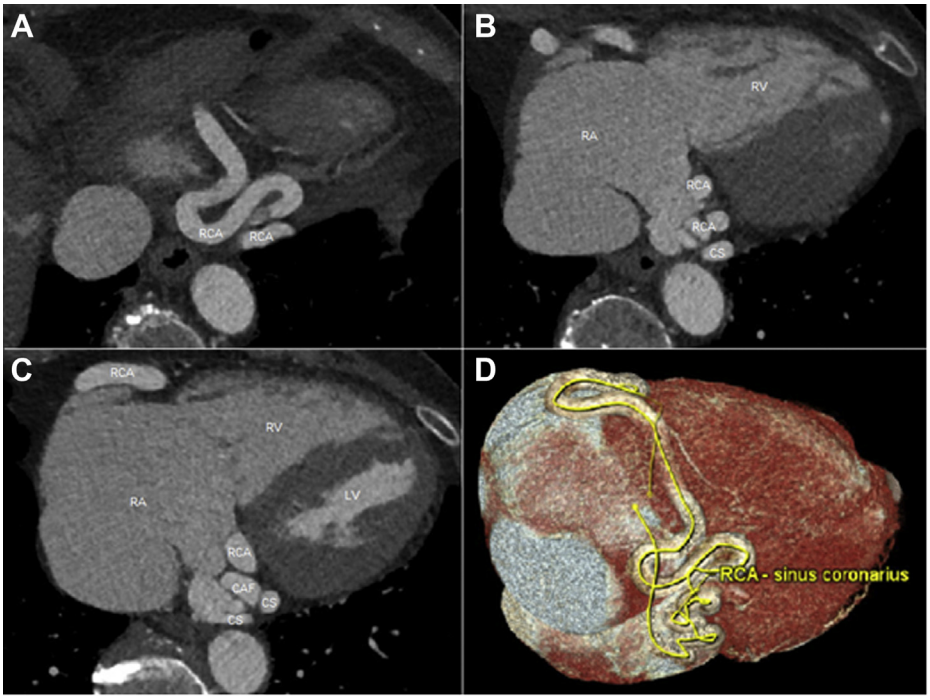
Laboratory results showed a significant elevation of 353 ng/l (normal value: <105 ng/l) brain natriuretic peptide concentration, and other findings were unremarkable. Transthoracic echocardiography visualized mild dilation of the left and severe dilation of the right atrium, moderate dysfunction of the left ventricle with an ejection fraction of 39%, severe dilation and dysfunction of the right ventricle, and severe tricuspid regurgitation. Furthermore, transthoracic echocardiogram revealed a large, aneurysmatic ostium of the right coronary artery (RCA) with a distal convolute, raising the suspicion of a fistula draining into the coronary sinus (CS). Computed tomography angiography visualized the presence of a severely dilated RCA fistula, draining into the CS, with dilation of the CS, right atrium, inferior vena cava, and liver veins (Figure 1).

MANAGEMENT

The patient experienced AF with high ventricular response rate and subsequent heart failure. After stabilization and rate control, she underwent heart catheterization. Coronary angiography showed a large and very tortuous RCA, with an offtake of the ostium of the ramus descendens posterior (RDP) and ramus posterolateralis (RPL) distal to several 180° turns. Thereafter, the fistula showed a tortuous network with diffuse exit of contrast into the CS. Hemodynamic measurements within the cardiac chambers appeared to be normal without pulmonary hypertension.

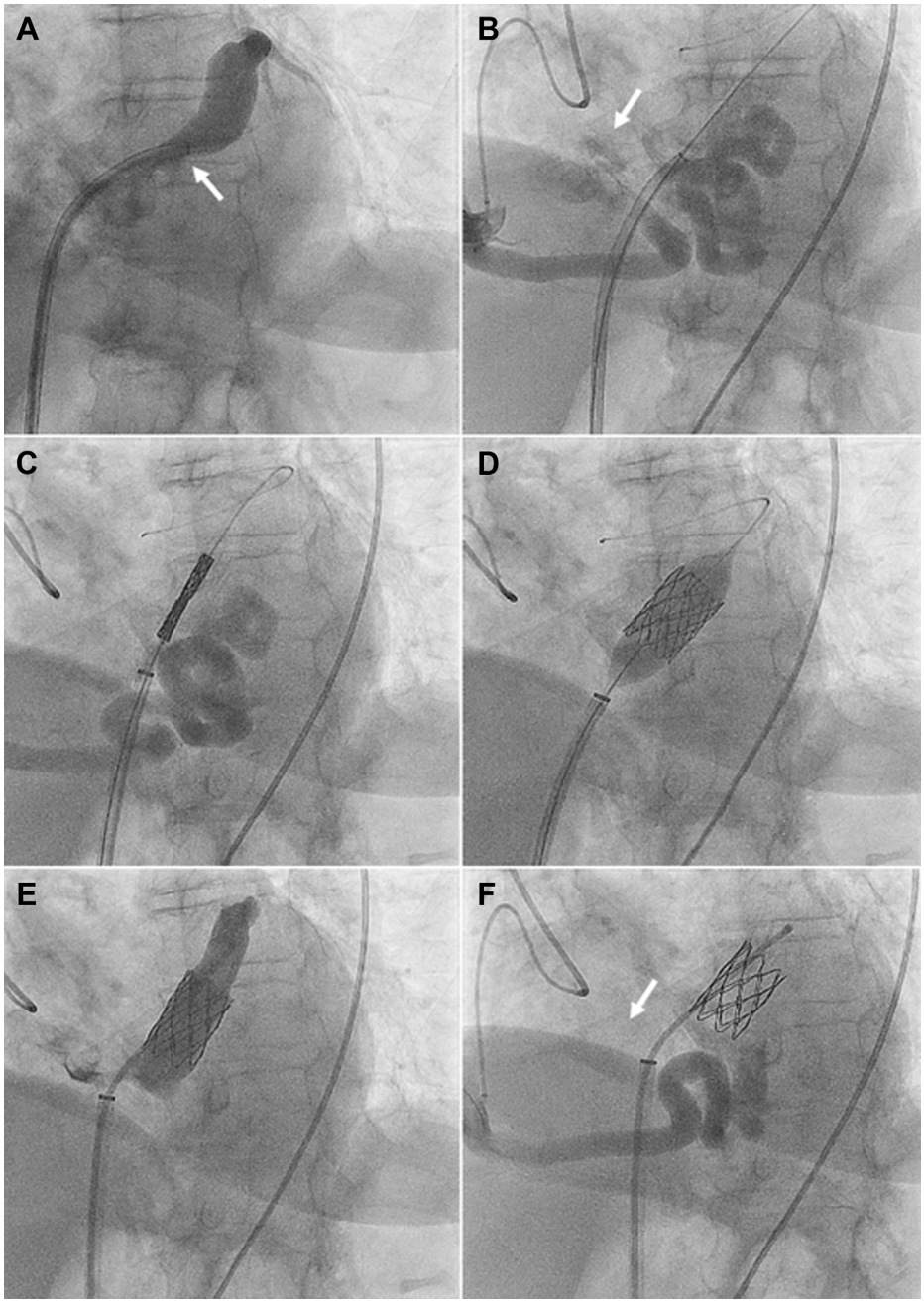
Initially, the adults with congenital heart disease (ACHD) heart team recommended surgical closure of the coronary fistula, bypass surgery with grafts on the RDP and RPL, and concomitant tricuspid annuloplasty. The patient received medical therapy to

FIGURE 1 Computed Tomography Angiography of the Heart



(A) Aneurysmatic tortuous right coronary artery. **(B and C)** The tortuous right coronary artery fistula drains into the coronary sinus and right atrium. **(D)** 3D image of the tortuous anatomy and connection between the right coronary artery fistula and coronary sinus into the right atrium. CAF = coronary artery fistula; CS = coronary sinus; LV = left ventricle; RA = right atrium; RCA = right coronary artery; RV = right ventricle.

FIGURE 2 Procedural Angiography



(A) Coronary sinus with compressed outlet into the right atrium **(arrow)** and dilated distal part. **(B)** tortuous right coronary artery with fistula draining into the coronary sinus, seen as contrast staining in the right atrium **(arrow)**. **(C and D)** Positioning and deployment of the covered CP stent in the coronary sinus. **(E)** Result of the covered CP stent with removal of the compressed outlet of the coronary sinus. **(F)** Result of the covered CP stent with closure of the fistula, seen as no contrast in the coronary sinus and no contrast staining in the right atrium **(arrow)**.

optimize her condition prior to the surgical intervention. Medical therapy consisted of rate control with beta blockers, diuretics, and an angiotensin-converting enzyme (ACE) inhibitor to treat the heart failure symptoms. Several weeks after the initiation of heart failure treatment and restoration of sinus rhythm, her clinical condition improved, and the transthoracic echocardiogram showed a significant

reduction of the tricuspid regurgitation. Without the need for tricuspid annuloplasty, transcatheter treatment options to close the fistula became more attractive and in a second ACHD heart team it was decided to perform an attempt to close the fistula percutaneously.

PERCUTANEOUS CLOSURE OF THE CORONARY FISTULA. An antegrade approach with closure of the distal RCA was not preferable because of the tortuosity of the RCA fistula (Video 1). Therefore, the percutaneous closure attempt was performed by retrograde approach; however, a approach through the CS, the fistula could not be engaged after multiple attempts. During the procedure a compression of the CS (Video 2), caused by convolutes of the fistula was seen, after which was decided to close the fistula with the implantation of a covered stent.

The closure was successfully performed by using a covered CP stent (28-mm NuMED, Inc). The covered stent was introduced through a 12-F right femoral vein guiding catheter, placed in the CS at the edge of the outlet of the CS into the right atrium, and deployed with a 16-mm balloon-in-balloon balloon (Videos 3 and 4). With the implantation of the covered CP stent, 2 issues were resolved: the compressed part of the CS was dilated, and the exit of the fistula between the RCA and the CS was sealed off and thus closed (**Figure 2**).

FOLLOW-UP

Two weeks after the procedure, the patient reported an improvement of her overall condition and did not experience any dyspnea or orthopnea. No complications were reported after 14 days of follow-up. One month after closure, a transthoracic echocardiogram was performed and showed proper positioning of the covered CP stent in the CS with a sustained closed fistula. The tricuspid regurgitation was further mildly reduced compared to pre-procedure. The patient reported an initial improvement of her overall condition, which still advances every day. After 4 months, the patient was still in sinus rhythm and symptom-free.

DISCUSSION

Coronary artery fistula (CAF) is a rare anomalous connection between a branch of the coronary arteries and one of the cardiac chambers or any other major blood vessel and has a large variation (1,2). The estimated overall incidence is 0.002%, and the incidence in adults undergoing coronary angiography is 0.13% to 0.6% (3,4). Most of these patients are asymptomatic, but the CAF bypasses the myocardial capillary network which could lead to myocardial ischemia due to coronary steal phenomenon and volume overload with clinical signs of heart failure and biventricular overload due to atrioventricular shunting (1-3). Other findings can be atrial or ventricular arrhythmia, aneurysmatic dilation of the involved vessels, rupture of the fistula, thrombosis, and infective endocarditis (1,3).

Treatment options are available for asymptomatic and symptomatic CAFs and include conservative treatment with medical therapy and surgical or transcatheter treatment options respectively (1,2,4). Patients with asymptomatic CAFs without signs of ventricular overload, ischemia, or arrhythmia can be treated conservatively. Symptomatic CAFS, however, are always considered for surgical or transcatheter treatment. The most important advantage of transcatheter closure over surgical closure is the avoidance of sternotomy/thoracotomy, cardiopulmonary bypass, and the procedure-related risks with additional complications, resulting in a lower recovery time and improved cosmetic outcome (1,4). Late complications are rare and beneficial long-term outcomes have shown the safety of transcatheter closure of CAFs; however, recanalization of the fistula can occur, and therefore long-term follow-up is recommended (2,3).

The anatomical pathway of CAFs varies and mostly originates from the RCA (55%), followed by the left coronary artery (35%), or both coronary arteries (5%), and drain into low-pressure structures, mainly into the right ventricle (41%), right atrium (26%), pulmonary artery (17%), coronary sinus (7%), left atrium (5%), left ventricle (3%), and superior vena cava (1%) (5). A CAF of the RCA draining into the CS is rare, and only a few case reports have been published (6). Most patients with a symptomatic CAF between the RCA and CS underwent surgical treatment (7).

To the authors' knowledge, the present case is the first to report a percutaneously closed fistula between the RCA and CS with a covered CP stent. The use of a covered stent to close fistulae is proven to be successful in different settings, such as arteriovenous fistulae, carotid-cavernous fistulae or secondary to percutaneous coronary interventions. In these settings the covered stent is deployed in the feeding artery to seal the entry port of the fistula. In our case the mechanism of closure is slightly different, the covered stent is placed in the drainage site (in this case the CS), from which the stent seals off and thus closes the fistula.

CONCLUSIONS

There is limited reported experience with the use of devices, including coils, occluders, plugs, and covered stents for transcatheter closure of CAFs. The closure technique in this report, deployment of the covered stent in the drainage site, shows the broad possibilities of transcatheter techniques to close fistulae and could contribute to further developments in the transcatheter closure spectrum of CAFs.

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KEY WORDS congenital heart defect, coronary circulation, coronary vessel anomaly, stents, treatment

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