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CASE REPORT

INTERMEDIATE

CLINICAL CASE

Sinus Node Dysfunction After Percutaneous Transcatheter Closure of Right Coronary Artery–Superior Vena Cava Fistula

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ABSTRACT

Congenital right coronary artery-superior vena cava (RCA-SVC) fistula is rare and typically does not manifest any symptoms until the fifth decade of life. The present case demonstrates a 48-year-old woman who developed Sinus node dysfunction of unknown cause after Percutaneous coil embolization of the RCA-SVC fistula requiring permanent pacemaker. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2023;16:101890) © 2023 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 48-year-old woman presented with palpitations. She denied experiencing any chest pain, diaphoresis, syncope, presyncope, shortness of breath, vision changes, confusion, tongue biting, or fatigue. The patient was afebrile and hemodynamically stable, with a blood pressure of 116/70 mm Hg, and a pulse rate of 66 beats/min. She had normal vesicular breath sounds and normal heart sounds, with no reported murmur or s3/s4 sounds.

LEARNING OBJECTIVES

- To understand the anatomy of the RCA fistula in computed tomography angiography and its potential association with sinus node dysfunction.
- To consider this occurrence when evaluating the risks and benefits of managing similar cases.

PAST MEDICAL HISTORY

The patient reported that she had a left knee surgery 23 years ago with no major complications thereafter. Additionally, she had a history of migraines with aura and occasional palpitations. She denied any history of thyroid disease, sleep apnea or sleep disordered breathing, chemotherapy, radiation, myocardial infarction, autoimmune disease, hypertension, congestive heart failure, bleeding, or telangiectasia.

DIFFERENTIAL DIAGNOSIS

Based on the patient's clinical symptoms and the findings of the echocardiography and the color Doppler, the differential diagnosis included patent foramen ovale, undiagnosed atrial septal defect,

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ABBREVIATIONS AND ACRONYMS

CAF = coronary artery fistula

RCA = right coronary artery

SVC = superior vena cava

anomalous pulmonary venous connections, or an unroofed coronary sinus.

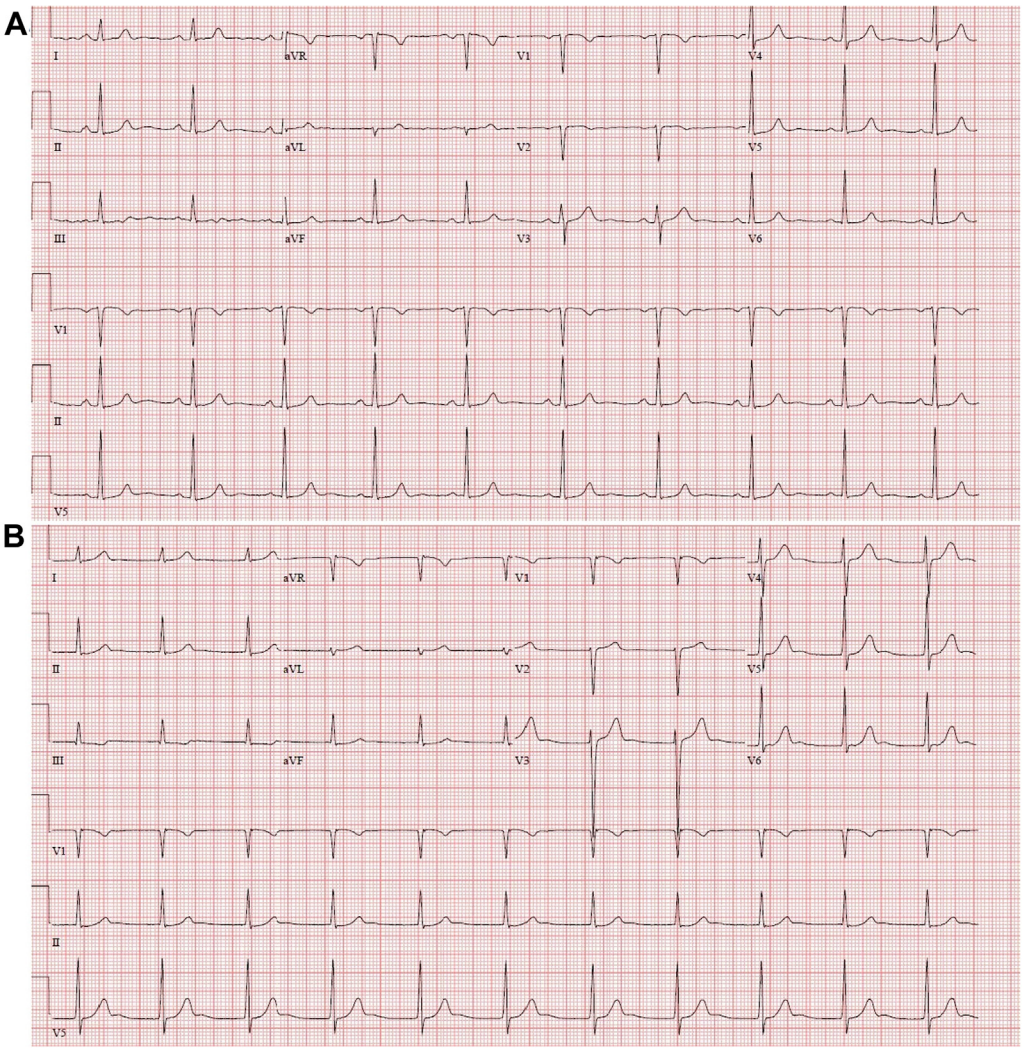
INVESTIGATIONS

The 12-lead electrocardiography and telemetry results showed normal sinus rhythm, as seen in **Figure 1A**. Transthoracic echocardiography revealed a moderately enlarged left ventricle with elevated filling pressure and a mildly enlarged right ventricle, with a right ventricular systolic pressure of 55 mm Hg. Color Doppler revealed a patent foramen ovale with bidirectional shunting, and significant right-to-left shunting was noted post-Valsalva maneuver.

Computed tomography angiography showed a dominant right coronary artery (RCA), which was enlarged proximally, giving rise to a sizable tortuous vessel coursing posteriorly and superiorly between the right atrium and aortic root, in or near the crista terminalis, finely coursing cranially and laterally, and entering the superior vena cava (SVC), as seen in **Figure 2**.

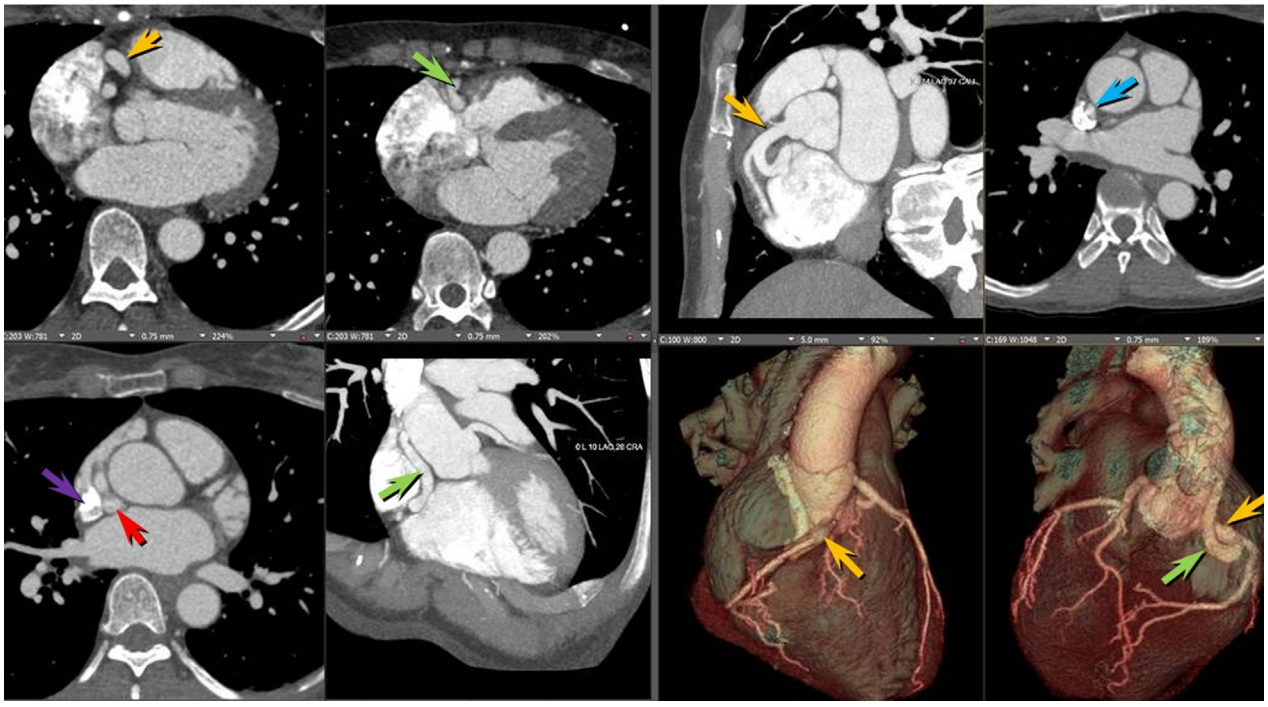
A subsequent right heart catheterization revealed an RCA-SVC fistula, as seen in **Figure 3** and Video 1,

FIGURE 1 **ECG Preprocedure and Postprocedure**



(A) Pre-procedure electrocardiogram (ECG). **(B)** Postprocedure ECG.

FIGURE 2 **Coronary CT Angiography**



Right coronary artery just beyond origin proximal to fistula **(orange arrow)**. Proximal fistula **(green arrow)**. More distal fistula near superior vena cava (SVC) (**red arrow**; SVC = **purple arrow**). **Blue arrow** is where fistula drains into SVC, at about the 2 or 3 o`clock position along the medial distal SVC posterior to the crista terminalis and a bit medial and caudal to the location.

with normal right heart pressures and no evidence of atherosclerotic coronary artery disease.

MANAGEMENT

After reviewing guidelines and discussing treatment options with the patient, a decision was made to perform percutaneous coil embolization of the RCA-SVC fistula. An 8-F JR4 catheter was used to cannulate RCA, and a 014 TRAVERSE guidewire was advanced into distal RCA. Then, a 014 TRAVERSE guidewire, and 130-cm Lantern microcatheter (Penumbra) were advanced into the fistula. In a sequential manner, Ruby coil 6 mm × 50 cm (Penumbra), two Ruby coils 8 mm × 60 cm (Penumbra), and POD Packing Coil j-Soft 60 cm (Penumbra) were used. Multiple views were obtained in between coil advancement to ensure no migration as seen in **Figure 4** and Video 2. After the first coil advancement, the patient's heart rate decreased to 50-60 beats/min, initially showing isorhythmic atrioventricular dissociation and then junctional rhythm. Despite the rhythm change, the QRS interval was narrow, and the patient was hemodynamically stable. Atropine was administered, which caused rate acceleration but no P waves.

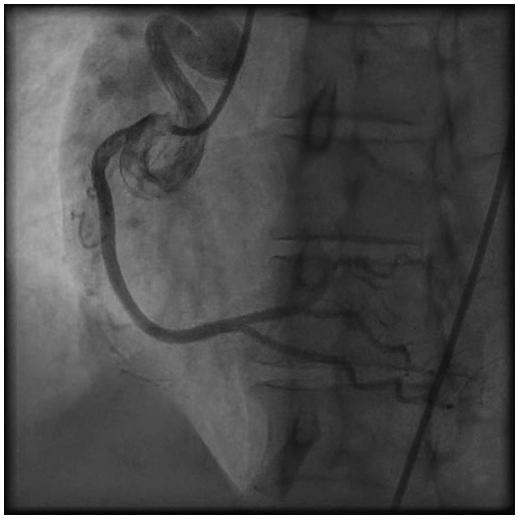
Following the procedure, the patient was hemodynamically stable with a stable escape rhythm as seen in **Figure 1B**. Initially, the decision was made for close observation, and the patient was discharged on apixaban. A week after the procedure, the patient reported a heart rate ranging between 40 and 60 beats/min, increasing up to 80 beats/min during exertion. She also noted fatigue but denied any palpitations, chest pain, shortness of breath, presyncope, or syncopal events. After a thorough discussion with the patient, it was decided to implant a single-chamber atrial pacemaker.

DISCUSSION

The case report raises 2 critical questions:

1. How was the medical decision made to close the coronary arteriovenous fistula?

FIGURE 3 RCA-SVC Fistula Before Coil Embolization



Fluoroscopic view of the right coronary artery–superior vena cava (RCA-SVC) fistula before coil embolization.

2. What could explain the persistent junctional rhythm after closure?

The management of asymptomatic coronary fistulas is still controversial; however, many experts agree that closure is necessary in cases where there is a significant shunt or aneurysmal dilatation. If left untreated, larger coronary artery fistulas (CAFs) can worsen the shunt and exacerbate symptoms over time. The presence of a significant shunt or aneurysmal dilatation justifies closure. Larger CAFs can enlarge over time. Diagnostic imaging plays a crucial role in identifying the origin, termination, and complexity of the fistula, as well as evaluating the dilation of the feeding artery. CTA is the preferred noninvasive imaging method for diagnosing CAF. Based on the morphology and origin of our patient's CAF, as shown on the computed tomography angiogram, it would be classified as Sakakibara type A.[1] The 2008 American College of Cardiology/American Heart Association guidelines state that CAFs that need to be evaluated for closure are the following: 1) large fistulas regardless of symptomatology; and 2) small to moderate fistulas that are linked to myocardial ischemia, arrhythmia, endarteritis, unexplained ventricular diastolic or systolic dysfunction, and ventricular dilation.[2]

Effective management of CAFs requires the evaluation of patient and CAF anatomy and a heart team approach involving interventional cardiologists, cardiothoracic surgeons, and those with training and expertise in congenital heart disease. Guideline-directed strategies for CAF closure include surgical and transcatheter techniques, with the latter requiring favorable coronary anatomy and successful cannulation of the artery of origin. Multiple catheter-based closure tools such as occlusion coils, detachable balloons, and deployable stents are available. It is crucial to address the distal vessel entry site before transcatheter embolization as this helps to prevent complications such as distal coil migration or incomplete shunt occlusion. Therefore, the preprocedural radiologic report should focus on delineating the distal entry site.[3]

The patient's bradycardia and junctional rhythm after CAF occlusion could be caused by one of the following. 1) Occlusion of the patient's sinoatrial node artery. 2) Mechanical compression during occluder expansion on the upper right cardiac and aortocaval ganglia leading to an increased parasympathetic activity, followed by sinus nodal inhibition. 3) Manifestation of Branham sign, which is mostly seen in patients undergoing hemodialysis. The presence of an arteriovenous fistula results in increased cardiac output caused by low resistance across the arteriovenous shunt. Occlusion of an arteriovenous fistula leads to an increase in arterial impedance and a decreased effective cardiac output because blood flow is redirected through the higher-resistance peripheral vasculature. A compensatory increase in cardiac contractility paradoxically activates baroreceptors and increases vagal tone. Thus, closure of an arteriovenous fistula results in a physiologic, bradycardic response.

A usual physiological response to the sudden closure of an arteriovenous fistula is a temporary bradycardia, which can be resolved with the administration of atropine or by elevating the lower extremities when lying down. However, in this case, the patient has been experiencing a persistent junctional escape rhythm after the closure of the coronary arteriovenous fistula, which is not a typical response.

FOLLOW-UP

During the subsequent follow-up visits, the patient did not report any symptoms, but she had a few brief episodes of fluttering that lasted between 5 and 10 seconds. Pacemaker interrogations revealed an atrial-

paced rhythm with a heart rate ranging from 60 to 150 beats/min and a pacing burden of 90%. Follow-up computed tomography angiography showed that the proximal RCA had decreased in size to 4 mm from the previous 6 mm, indicating successful occlusion of the fistulous connection.

CONCLUSIONS

It is crucial to treat certain CAFs to avoid complications such as arrhythmias, thrombosis, and volume overload. To prevent the need for a pacemaker implantation, it is important to estimate the risk of sinus dysfunction before the occlusion procedure. This can be done by conducting temporal occlusion testing or retrieving the coil if signs of decreased heart rate emerge during the first occlusion. Knowledge of the occurrence of sinus dysfunction is helpful in making medical decisions for managing patients with coronary arteriovenous fistula. An individualized approach is required for transcatheter closure of the RCA fistula, considering the risks and benefits, including long-term complications and potential sinus node dysfunction.

FIGURE 4 RCA-SVC Fistula After Coil Embolization

Fluoroscopic view of the right coronary artery–superior vena cava (RCA-SVC) fistula after coil embolization.

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KEY WORDS embolization, fistula, junctional rhythm, palpitations, sinus node

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