Occlusion of a congenital right coronary–vena cava superior fistula induces temporary junctional bradycardia and atrial fibrillation

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Introduction
The finding of a congenital right coronary–vena cava superior (RCVCS) fistula is extremely rare. The overall prevalence of coronary artery fistulae is 0.02%–0.9%,1,2 of which the coronary-to-pulmonary artery fistula is the most common type (78%) and the coronary-to-vena cava superior fistula the least common (1.8%). Symptoms such as dizziness, palpitations, angina, reduced exercise capacity, and/or fatigue may accompany this phenomenon, but most coronary fistulae are asymptomatic.

Patients who developed secondary pulmonary hypertension owing to left-to-right shunting via the arteriovenous fistula will likely be symptomatic, as illustrated in this case report, whereas in normal-pressure fistulas symptoms are less frequent.3 Therapeutic occlusion of the fistula is, certainly in the case of symptoms, recommended, since an increased risk exists for endocarditis, transient ischemic attacks, coronary steal phenomenon, pericardial tamponade, myocardial ischemia, and even hydrops fetalis when left untreated.4–6

Various treatment strategies are described, varying from cardiothoracic surgery to minimal invasive occlusion by using devices like Amplatzer plugs and Gianturco coils.7 Owing to its rarity, tailor-made therapy differs per center and depends on available expertise. Interestingly, to our knowledge this is the first case report describing the relation of closing the fistula and triggering of arrhythmia, and raises the question of a potential relation between paroxysmal atrial fibrillation (AF) and a coronary fistula.

Case presentation
A 63-year-old woman presented with dizziness and fatigue of a few months’ duration. Other than a bilateral cataract operation, her past medical history was unremarkable. No congenital abnormalities were reported in her family. A Holter test revealed paroxysmal AF. After failure of antiarrhythmic drugs, the patient opted to undergo pulmonary vein isolation (PVI). An adjacent diagnostic coronary catheterization showed a large RCVCS fistula (Figure 1A) with Qp: Qs of 1.83, next to significant stenoses of the mid left anterior descendens and first diagonal artery. The measured hemodynamic parameters (all pressure values expressed in mm Hg) were as follows: cardiac output 7.2 L/min (N: 4–8), aorta pressure 149/58 (N: 100–140/60–90), right atrium 11 (N: 2–6), right ventricle 41/9 (N: 15–30/2–8), pulmonary wedge 11 (N: 9–18), pulmonary artery 35/10 (N: 15–30/8–15). A transesophageal echocardiogram and computed tomography of the thorax revealed a large communicating fistula originating from the right proximal coronary to the vena cava superior (Figure 2A and B), confirming a slight increased right ventricular pressure and mean pulmonary pressure without tricuspid regurgitation. After assessment by a multidisciplinary team of an electrophysiologist, an interventional cardiologist, and a thoracic surgeon, a successful occlusion with a 12 mm Amplatz device prior to percutaneous coronary intervention and PVI was performed (Figure 1B). A clear increase of contrast-filled collaterals from the right coronary artery was seen after closure. Immediately after fistula closure a profound bradycardia occurred with a junctional escape rhythm of 40 beats/min (Figure 3A) lasting for 20 minutes, followed by paroxysmal AF (Figure 3B), which converted spontaneously after 13 hours. Volume supplmentation was sufficient to treat the associated hypotension adequately. The patient remained hemodynamically stable after additional percutaneous coronary intervention of the left anterior descendens and first diagonal artery. Planned PVI was canceled pending the response of the occlusion of the fistula on the cardiac rhythm. During a follow-up time of 11 months, no AF recurrence occurred.

KEYWORDS Congenital right coronary-to-vena cava superior fistula; RCVCS; Junctional bradycardia; Paroxysmal atrial fibrillation; Amplatzer plug; Bezold-Jarisch reflex (Heart Rhythm Case Reports 2016;2:169–172)

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Discussion

This case report describes a unique case of an RCVCS fistula in a patient with paroxysmal AF. Immediately after fistula closure, a profound bradycardia consisting of a junctional escape rhythm occurred, followed by an episode of paroxysmal AF (Figure 3A and B). Two intriguing questions arise from this case report: (1) How to explain the transient rhythmological findings after closure?, and (2) Is there any relation between paroxysmal AF and the existence of an RCVCS fistula?

The most probable mechanism to explain question 1 is that the sudden hypovolemia after closure of the RCVCS fistula triggered a Bezold-Jarisch reflex via the muscarine nonmyelinated afferent barosensor receptors in the atria (M5) and ventricles (M3), followed by an increased parasympatic inhibition of the sinoatrial node and atrioventricular conduction via the C-efferent fibers. Another potential mechanism is mechanical compression during Amplatzer expansion on the upper right cardiac and aortocaval ganglia, leading to an increased parasympathetic activity followed by sinus nodal inhibition. Also, a sudden hyperemia of the right coronary myocardial area (as shown in Figure 1B by more contrast-filled collaterals) could alternate the blood flow in the sinus and atrioventricular nodal artery. This sudden increased coronary flow may enhance myocardial acidosis owing to an increased myocardial oxygen demand on the basis of mechanoenergetic disturbance, which can be followed by the Bezold-Jarisch reflex. Other, less probable mechanisms are central mediated vagal overdrive due to prostaglandin, endothelin, or A2A antagonist release in response to fistula occlusion, whether or not synergized with propofol-induced hypovolemia, leading to the Bezold-Jarisch reflex. All these mechanisms would be expected to be temporary, and therefore do not fully explain the duration of the secondary arrhythmias.

Regarding the second question, could the fistula be a trigger of paroxysmal AF before closure or is it an epiphenomenon? The maintenance of sinus rhythm, almost 1 year after closure, suggests that there could be a link. Potentially the higher right atrial pressure could lead to atrial stretch, which in itself is a proven mechanism for focal firing of excitable regions. Also, the inverse relation between atrial stretch and autonomic inhibition lowering AF vulnerability via atrial autonomic ganglia is well described. Closure of the RCVCS fistula has the potential to stop the vicious circle of atrial dilation due to increased atrial stretch as a prerequisite for the domestication of AF. Although suggested by the disappearance of AF, whether closure of the fistula with presumed reverse remodeling of the atria is sufficient to maintain sinus rhythm over time remains to be seen. If AF were to recur, it could very well be that potentially other, more common AF initiating mechanisms, such as excitable pulmonary foci, were already present from the start or developed over time, further limiting a causal relationship between AF and a coronary fistula. Therefore, although a relation between paroxysmal AF and the RCVCS fistula in this case is suggested, the role of atriovenous fistula in the development of AF remains speculative and needs to be further studied.

Figure 1  A: An X-AP (anteroposterior radiograph) view showing clearly a right coronary–to–vena cava superior fistula (RCVCS). B: An X-cranial (cranial radiograph) view showing the successfully deployed Amplatzer plug (arrows) in the proximal sac that occludes the fistula. Note the increased collaterals from the right coronary artery (RCA) (asterisks). VCS = vena cava superior.
Conclusion
This case report highlights the potential occurrence of profound bradycardia and paroxysmal AF after RCVCS fistula closure. Since the patient had paroxysmal AF before closure and was free of AF after closure, a potential causal relation could exist between paroxysmal AF and RCVCS fistula. However, this remains to be proven.

Figure 2  A: A transesophageal short-axis view at 140 degrees, showing the large aneurysmatic sac at the proximal right coronary artery and a part of the right coronary–to–vena cava superior (RCVCS) fistula. B: A transversal coupé of computed tomography of the thorax with contrast, showing the large right coronary artery (RCA) to vena cava superior (VCS) fistula (arrows).

Figure 3  Post–right coronary–to–vena cava superior arrhythmia sequence after closure. A: First, a junctional escape rhythm of 40 beats/min lasting for 20 minutes, followed by B: paroxysmal atrial fibrillation converting spontaneously after 13 hours.
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