

## Loops Around the Heart – A Giant Snake-like Right Coronary Artery Ectasia with Unique Convolute and Fistula to the Coronary Sinus

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### Keywords:

Coronary artery ectasia and aneurysm, arterio-venous fistula; Intracoronary injection of contrast agent

### Key Clinical Message:

Intracoronary injection and simultaneous echocardiographic visualization of contrast agent (Sonovue) is a helpful method to detect fistula of coronary artery in to great vessel or chamber

## 1. Abstract

Coronary artery dilatation is an uncommon finding and is incidentally found during diagnostic coronary angiography or at necropsy. The pathogenesis of dilatation of coronary arteries is still not very well understood and therapeutic strategies are not clear. It is useful to know the difference between aneurysm and ectasia. In this report we demonstrate the diagnostic workup of an asymptomatic patient with a remarkable snake-like dilatation of the right coronary artery with unique convolute. For the first time we used intracoronary injection and simultaneous echocardiographic visualization of contrast agent (Sonovue) to proof a fistula to the coronary sinus. Like our patient, most of the patients are asymptomatic in absence of coronary artery disease and we decided on a conservative approach because of his very complex anatomy.

## 2. Introduction

Dilatation of coronary arteries is well described in many reports. The pathogenesis of this condition is still not very well understood and therapeutic strategies are not clear in asymptomatic patients and specially in absence of coronary artery disease. In this report we want to describe the case of an asymptomatic patient with a remarkable giant and snake-like dilatation of the right coronary artery ending in a unique convolute and arterio-venous fistula to the

coronary sinus. Furthermore we want to describe for the first time the Intracoronary injection and simultaneous echocardiographic visualization of sulphur hexafluoride contrast agent (Sonovue) as a very helpful method to detect fistula of coronary artery in to great vessel or cardiac chamber. We would like to take this opportunity to explain the difference between aneurysm and ectasia, as well as to describe the epidemiology, etiology, the course of the disease and treatment options. To our knowledge, there are no previous reports in the literature on such an extreme case with use of intracoronary injection and simultaneous echocardiographic visualization of contrast agent (Sonovue).

## 3. Case Report

A 65-year-old previously healthy patient was brought to the emergency department in January 2022 because of a sudden tachycardia during his bicycle training session, which he was able to record well with his Apple Watch. He regularly goes on intensive bike tours and so far, he has not noticed any cardiac problems or reduced performance. A new atrial flutter with slow, irregular conduction and normal ventricular rates were found in the emergency room. The clinical examination was normal. The laboratory examination revealed a discrete latent hypothyroidism as well as borderline increased high sensitivity troponin T values (20.9 ng/l) without dynamic changes. An oral anticoagulation was established

at a CHA2DS2-VASc score of 1 and the patient spontaneously converted to sinus rhythm a few days later. On sinus rhythm the 12-lead ECG showed a first-degree AV-block and minimal ST-depressions in V5 and V6, otherwise there was no evidence of ischemia or arrhythmias. We did a Stress-ECG and saw a very good physical condition with 170% of the expected performance. The patient did not show any symptoms, the circulatory behavior was normal and there was no increase of the ST-depression that already existed at rest.

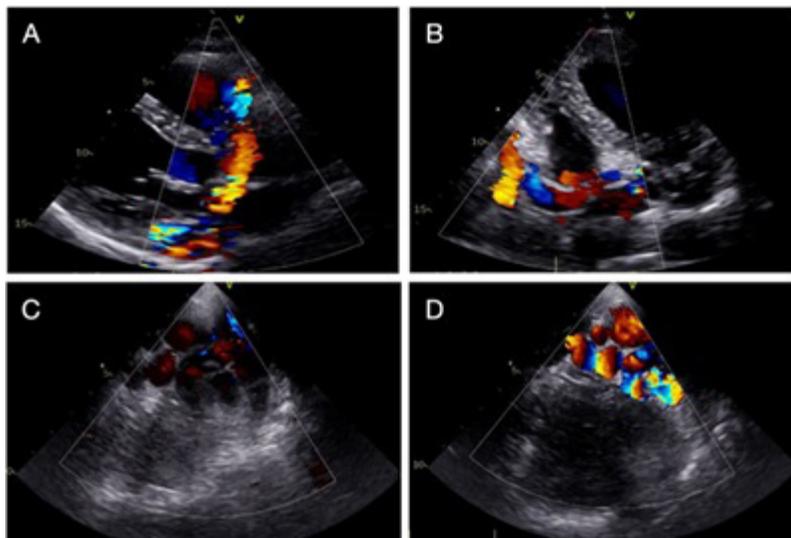
Apart from a clearly dilated right atrium (28cm<sup>2</sup>), no other relevant structural cardiopathy was found in the transthoracic echocardiography. However, an unclear color-doppler signal was detectable in several places around the heart and especially behind the left atrium. To clarify this further, a transesophageal echocardiography was performed. There we saw a sign of multiple tortuous vessels around the heart and an unclear convolute of vessels in the area behind the left atrium. Even after intravenous injection of lung-permeable (Sonovue) and non-lung-permeable contrast agent (agitated NaCl-bubbles), this finding could not be clarified (Figure 1).

To further clarify the anatomy, we performed a computed tomography angiography. A massively ectatic Right Coronary Artery (RCA) was found with a diameter of 17 mm and tortuous course

far into the periphery, with a parasagittal convolute between the aorta and the left atrium. No other concomitant anomalies were detectable (Figure 2 and 3).

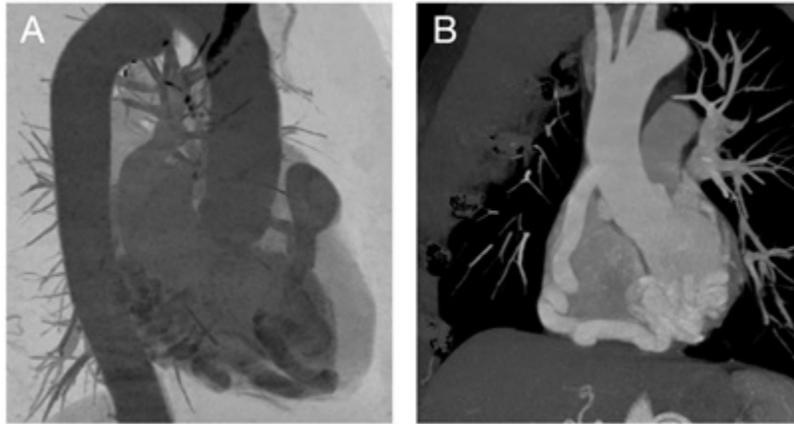
Since it was unclear where the convolute of the RCA ends, a coronary angiography was performed. There, normal left-sided coronary arteries were visualized, the RCA was grotesquely ectatic (Figure 4). To find out where the convolute of the RCA ends, we decided to inject contrast agent (Sonovue) into the RCA while performing echocardiography. To our knowledge, there is no reports about intracoronary injection and simultaneous echocardiographic visualization of contrast agent (Sonovue). Immediately after the injection we could clearly see the contrast agent in the coronary sinus just before it joins the right atrium (Figure 5). So we could prove a fistula of the convolute of the RCA into the coronary sinus. This arterio-venous fistula explains the echocardiographic findings of the significantly dilated right atrium. In the right heart catheter, however, no relevant systemic left-right shunt was found and all pressure values in the systemic and pulmonary circulation were normal.

Since the patient is completely asymptomatic and because of this complex anatomy, we decided on a conservative approach with a regular follow-up.



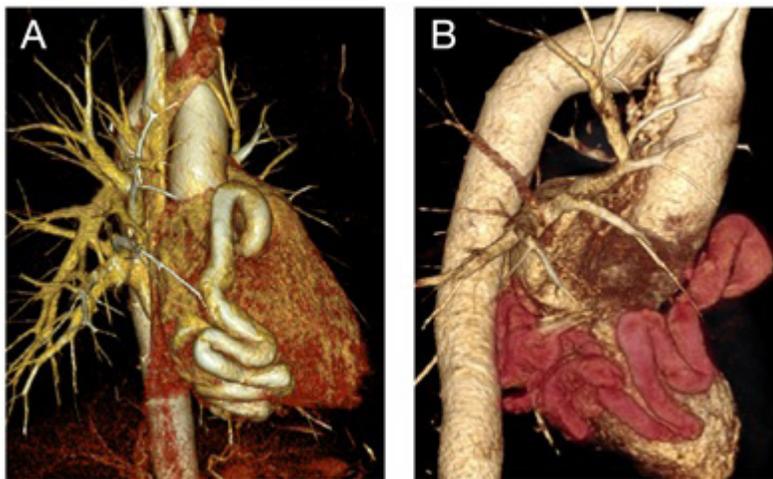
**Figure 1:** Transthoracic (TTE) and transesophageal echocardiography (TEE)

TTE with multiple color-doppler signals detectable in several places around the heart. A) Inflow from coronary sinus and probably the flow of the RCA. B) Flow around the base of the right ventricle and into the coronary sinus. C) Convolute of vessels in the area of the aorta and the left atrium. D) Blood flow within the convolute of vessels around the heart.



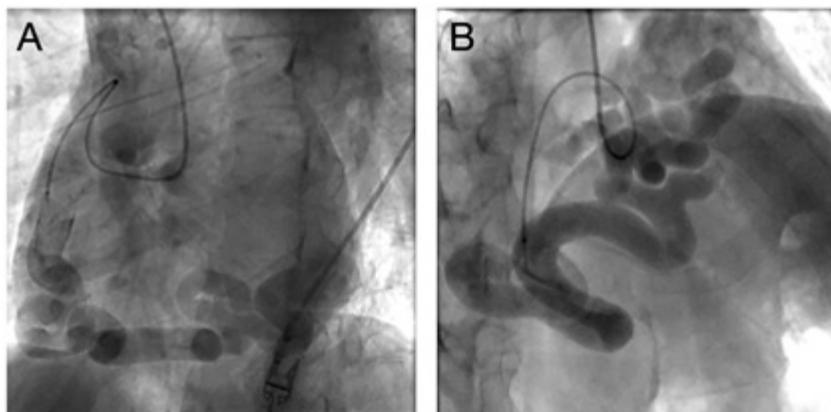
**Figure 2:** Coronary CT-angiography

A) Ectatic RCA with tortuous course and a parasagittal convolute of vessels between the aorta and the left atrium. B) Another view showing the convolute of the RCA behind the left atrium.



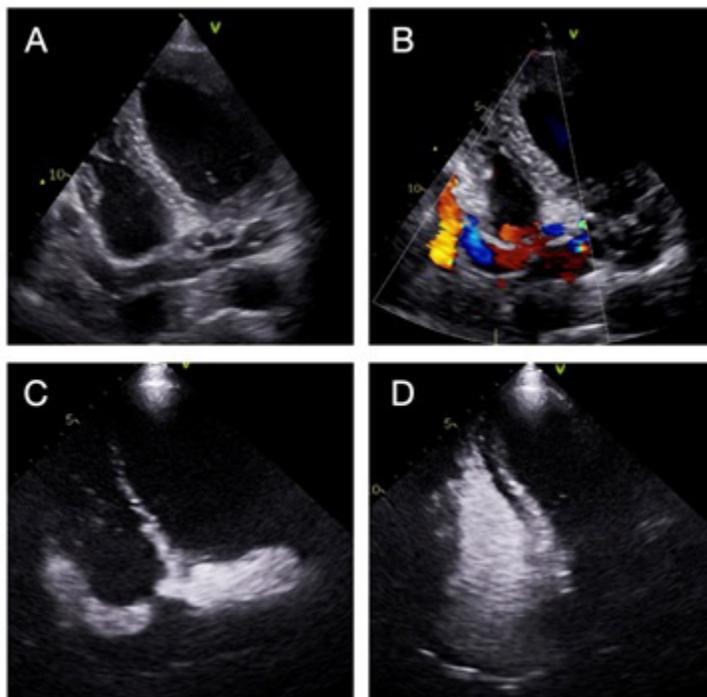
**Figure 3:** Computed tomography of the heart with 4D angiography

A) Reconstruction of the heart and aorta and the ectatic RCA with tortuous course. B) Reconstruction with the focus on the convolute of the RCA between left atrium and aorta



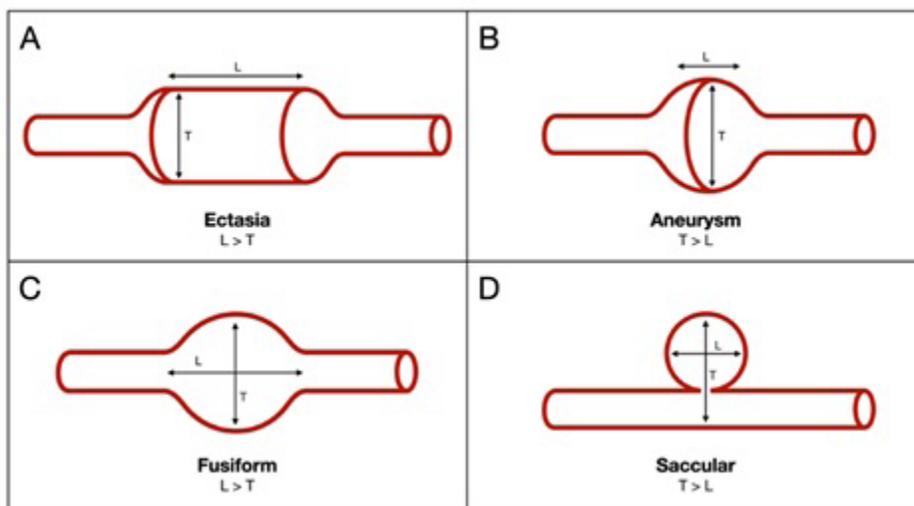
**Figure 4:** Coronary angiography of the right coronary artery

A) Grotesquely ectatic RCA with tortuous course. B) Convolute of the RCA ending in the coronary sinus.



**Figure 5:** Intracoronary injection and simultaneous echocardiographic visualization of the contrast agent (Sonovue) showing a fistula in to the coronary sinus

A) TTE shows a part of the RCA and the coronary sinus for orientation. B) TTE shows color doppler flow in the epicardial convolute, the coronary sinus, and the RCA. C) TTE shows the contrast agent in the RCA and the coronary sinus immediately after the intracoronary injection to the proximal part of the RCA. D) Visualization of the contrast in the right atrium and right ventricle after exiting the coronary sinus.



**Figure 6:** Difference between ectasia and aneurysm of coronary arteries

A) Ectasia: Increase of the diameter greater than 1.5 times of the segment of the normal adjacent artery with length > width. B) Aneurysm: focal increase of the diameter more than 1.5 to 2 times of the normal adjacent segment. C) Fusiform aneurysm with a longitudinal dimension > 200% of its transverse dimension. D) Saccular aneurysm with a transverse dimension greater than the longitudinal dimension.

**4. Discussion**

It is important to know the difference between coronary artery ectasia and aneurysm, although they may be a manifestation of the same pathological process [1]. Usually, the term ectasia is reserved to a diffuse dilatation of a coronary artery, and an aneurysm is a focal dilatation of the vessels [2].

There are many definitions of coronary aneurysms, but the most accepted is the focal increase of the diameter more than 1.5-2 <http://www.acmcasereport.com/>

times of the normal adjacent segment [3]. Coronary artery aneurysms are classified on the basis of the composition of the wall, the shape and the size of the vessel (Table 1). First, coronary artery aneurysms can be either “true” aneurysms or “false” aneurysms (pseudoaneurysms), a distinction that is based on the number of layers in the vessel wall [4]. The vessel wall in true aneurysms is composed of all three layers, whereas in pseudoaneurysms there is a loss of the wall integrity as a result of disruption of the external

elastic membrane for example as a result of coronary interventions [4]. Second, coronary artery aneurysms can be classified as saccular or fusiform on the basis of their shape. The saccular aneurysm has a transverse dimension greater than the longitudinal dimension of the segment, while the fusiform aneurysm has a longitudinal dimension  $> 200\%$  of its transverse dimension [5]. Third, on the basis of their size, some coronary artery aneurysms are called giant coronary artery aneurysms when the maximal diameter exceeds 20mm in adults or 8mm in children [6, 7].

Coronary artery ectasia is diagnosed when a segment of the artery is greater than 1.5 times the diameter of the segment of the normal adjacent artery with length  $>$  width [8]. It has been classified by Markis into four types depending of the distribution with the worst outcomes in types I and II because affecting more than one vessel [9]. Table 1 shows the classification and Figure 6 shows schematically the difference between ectasia and aneurysm of coronary arteries.

In this patient, the ectasia can be classified as type III based on the distribution pattern. However, this only refers to the proximal course of the RCA. The tortuous course and the convolute cannot be assigned to any classification. The drain into the coronary sinus allows this anomaly to be described as an arteriovenous fistula.

The reported frequency of coronary artery aneurysms varies widely from 0.3% to 5% because of varied angiographic criteria used to define coronary aneurysms with higher incidence in men than in women (2.2% vs 0.5%) [9-14]. Coronary artery ectasia is more common than coronary artery aneurysm [15]. The RCA is the most frequently involved vessel (40%–61%), which matches with our case, followed by the LAD (15%–32%) and the RCX (15%–23%) [9, 18, 19].

The etiology of coronary artery dilatation varies with the geographic location and the age of the patients. Atherosclerotic aneurysms and are most common in western countries and usually caused by poststenotic dilatation (50%), followed by congenital (17%) and infectious causes (10%), a special situation is found in Japan with Kawasaki disease as the predominant cause of coronary artery aneurysm. Vasculitides (other than Kawasaki), connective tissue disorders and collagenopathies are very rare causes of coronary artery dilatation [16-18]. The precise mechanism is not well understood, but the underlying histologic changes seem to be equivalent to the changes observed in atherosclerotic lesions [9].

Most of the reported cases of congenital aneurysms and ectasia are based on compensatory dilatation and associated with coronary fistulas as a connection between a coronary artery and a cardiac chamber or great vessel [20, 21]. Fistulas originate from the RCA in 52% of cases, followed by the LAD (30%) and the RCX (18%) [22]. In 90% of the patients, the drainage is to the right cardiac chambers with volume overload to the pulmonary vascular bed [23]. Since our patient has no coronary artery disease, a congenital ectasia based on a fistula that could possibly have caused this com-

pensatory dilatation is highly likely. However, this tortuous course and the massive convolute cannot be explained, especially with normal pressure conditions measured invasively in the systemic and pulmonary circulation.

Most of the patients remain asymptomatic, and aneurysms are incidentally found during diagnostic coronary angiography or at necropsy [2]. When patients are symptomatic, the clinical manifestations depend on the underlying cause and the complications of the dilatation. The natural outcome of atherosclerotic aneurysms is directly related to the severity of the coexisting obstructive coronary artery disease although no difference in mortality between patients with or without coronary artery disease was demonstrated [28, 29]. Rupture of aneurysms seems to be very unpredictable.

Gold standard in diagnosing coronary ectasia is coronary angiogram and Intravascular Ultra Sound (IVUS) is the best tool for assessing luminal size and determining arterial wall changes [30, 31]. Noninvasive technologies such as computed tomography and magnetic resonance coronary angiography are also used to diagnose coronary dilatation and may increase the rate of recognition because of increased availability of this technologies [32, 33].

Medical management depends on the underlying disease. Especially with a coexisting obstructive coronary artery disease, the treatment is performed by the underlying coronary artery stenosis. In the absence of the obstructive coronary artery disease, treatment is not well established because of the rarity of this condition [17, 19]. Conservative medical therapy aims to prevent thromboembolic complications with anticoagulant therapy and administration of antiplatelet drugs [24]. Calcium channel blockers prevent spasm of the coronary arteries and the use of beta blockers should be avoided, because they can lead to spasm of the coronary arteries due to an overlap of the alpha receptor [1, 5, 9]. Both pharmacological and interventional methods can be used in the case of arterial thrombotic occlusion [34]. Since there are numerous individual variants of ectasia, there is no defined surgical therapy and the individual benefit-risk ratio should be calculated very carefully. A surgery may be considered to avoid complications and is indicated in patients with obstructive coronary artery disease or evidence of embolization leading to myocardial ischemia and also in those patients with very large dilatation with increased risk of rupture [25]. Percutaneous treatment with stents is another option in patients with a fistula that needs closure [26, 27].

As already mentioned, we chose a conservative therapy in our case because of the complexity of the anomaly, normal invasive pressure conditions and the patient's absolute lack of symptoms. While the patient is already on oral anticoagulation as a consequence of atrial flutter and a CHA2DS2-VASc score of 1 point, there is also protection against thromboembolic events as a possible complication of ectasia. Just recently, an article with a similar anomaly was published [35]. However, the course of the RCA was less spectacular than in our case and the use of intracoronary injection and

simultaneous echocardiographic visualization of contrast agent (Sonovue) was required to visualize the fistula in our patient. This approach was used for the first time. Due to the higher shunt vol-

ume and the patient's symptoms in the other case report, a surgical therapy was necessary, as opposed to ours.

**Table 1:** Classification of coronary artery dilatation

Coronary Artery Dilatation	Description
<b>Aneurysm:</b>	Focal Dilatation
<b>Vessel Wall Composition:</b>	
True Aneurysm	Wall with three layers: adventitia, media and intima
False Aneurysm	Wall with only one or two layers
<b>Shape:</b>	
Saccular Aneurysm	Diameter transverse > longitudinal
Fusiform Aneurysm	Diameter transverse < longitudinal
<b>Size:</b>	
Giant Aneurysm Adult	>20mm in diameter
Giant Aneurysm Children	>8mm in diameter
<b>Ectasia</b>	Diffuse Dilatation
Type I	Diffuse ectasia in two or three vessels
Type II	Diffuse ectasia in one vessel and localized disease (aneurysm) in other
Type III	Diffuse ectasia in only one vessel
Type IV	Coronary aneurysm in one vessel

## 5. Conclusion

Coronary artery dilatation is an uncommon finding and is incidentally found during diagnostic coronary angiography or at necropsy. However, with the advent of noninvasive methods for the evaluation of the coronary arteries, the dilatation is more frequently found. It is useful to know the difference between aneurysm and ectasia. In our case we could see the first signs in the echocardiography and could proof a remarkable giant snakelike right coronary artery ectasia with unique convolute in the coronary CT angiography and coronary angiography. The drain into the coronary sinus allows this anomaly to be described as an arteriovenous fistula. Intracoronary injection and simultaneous echocardiographic visualization of contrast agent (Sonovue) is used the first time in our case and could be very helpful to detect fistula of coronary artery in to great vessel or cardiac chamber. Like our patient, most of the patients are asymptomatic in absence of coronary artery disease, which is the most common cause of coronary artery dilatation in western countries and in adults. The clinical manifestations and the medical management depend on the underlying disease and the complications of the dilatation such like thromboembolic events and ischemia.

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