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An unusual pair: coronary artery fistula and coronary sinus ostium stenosis as a cause of refractory angina

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Background Coronary fistula are rare and often present in early adulthood with symptoms of right heart overload from left to right shunting or ischaemia in the distal coronary bed due to coronary steal.

Case summary A 73-year-old lady with prior history of supraventricular tachycardia, dyslipidemia and a right coronary artery (RCA) to coronary sinus (CS) fistula, presented with progressive angina. She did not have evidence of ischaemia in the RCA territory on nuclear imaging, and cardiac computed tomography (CT) did not show coronary artery disease but revealed a significantly dilated CS and coronary venous tree. She was found to have CS ostial stenosis and, under transesophageal echocardiographic guidance, underwent successful balloon angioplasty of the CS ostium, with decompression of the coronary venous circulation and resolution of her angina.

Discussion Coronary fistula draining to the CS are rare, and association with CS ostial stenosis has been reported very infrequently. CS ostial stenosis can cause elevated coronary venous pressure, leading to decreased global coronary perfusion and symptoms of angina or heart failure. Previous case reports of coronary fistula and CS ostial stenosis were treated with either medical therapy or surgery, and our case is the first to our knowledge to report successful percutaneous treatment.

Keywords Coronary sinus stenosis • Coronary fistula • Structural interventions • Angina • Balloon angioplasty

ESC Curriculum 2.2 Echocardiography • 2.4 Cardiac computed tomography • 3.1 Coronary artery disease • 3.4 Coronary angiography • 9.7 Adult congenital heart disease

Learning points

- Coronary artery fistulae are rare and often asymptomatic; when the left-to-right shunt is significant, they may cause heart failure symptoms.
- Coronary sinus ostial stenosis is associated with increased coronary venous pressure that may lead to decreased coronary perfusion and symptoms of angina.
- Relief of coronary sinus stenosis can be achieved percutaneously.

Introduction

Isolated coronary arteriovenous fistulas are uncommon, with an estimated incidence of 0.002% to in the general population,[1] though they are more commonly diagnosed as cross-sectional imaging is becoming more common. Adults with coronary fistulas present after a murmur is heard on exam or with shortness of breath, fatigue, exercise intolerance or angina. They may be isolated or associated with other congenital heart defects; percutaneous or interventional treatment is indicated for symptoms attributable to a significant left to right shunt, or ischaemia, and a moderate or large fistula.[2,3]

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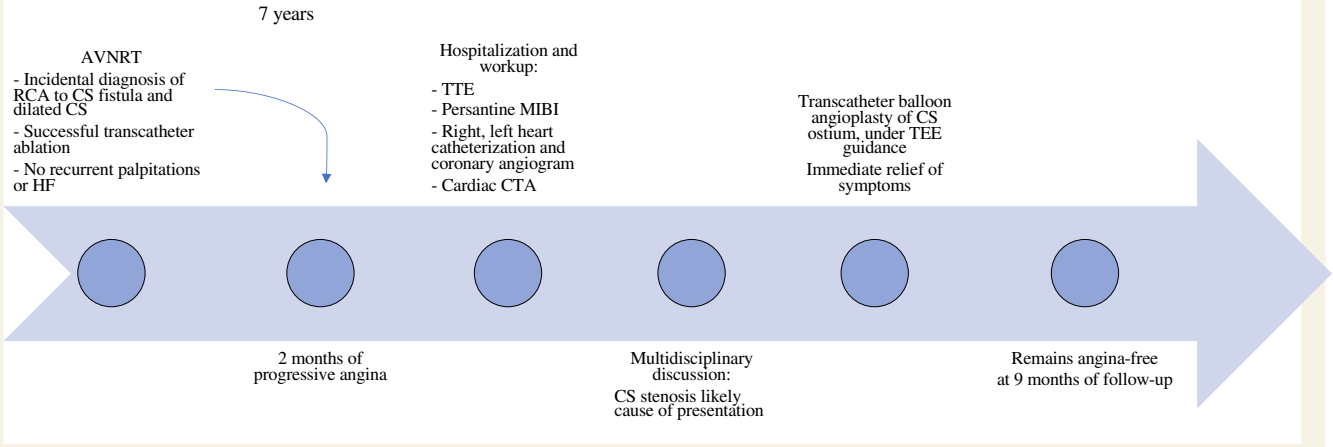
Importantly, aneurysmal segments of the coronaries proximal to the fistula often remain dilated with abnormal flow, and the risk of coronary thrombosis may increase after occlusion of the fistula.[4]

Timeline



Case presentation

A 73-year-old female presented to her local hospital with a 2-month history of worsening exertional dyspnoea and chest tightness. She did not report any presyncope, syncope or leg swelling. Her past medical history was significant for dyslipidemia, atrioventricular nodal reentry tachycardia which had been successfully ablated and asthma. She had a previous incidentally diagnosed right coronary artery (RCA) to coronary sinus (CS) fistula. She was on no cardiovascular medications.

Her cardiac examinations was significant for an elevated jugular venous pressure, and a grade II of VI holosystolic murmur heard best at the right and left lower sternal borders. Electrocardiogram showed normal sinus rhythm without evidence of ischaemia, and chest radiograph revealed a mildly enlarged cardiac silhouette. Her troponin I and N-terminal pro B-type natriuretic peptide were within normal parameters. A persantine myocardial perfusion imaging did not reveal any inducible ischaemia. A coronary angiogram confirmed there was no significant coronary artherosclerosis. She continued to have angina with minimal effort. A repeat echocardiogram identified a small pericardical effusion and high-velocity jet at the outflow of the CS, suggestive of ostial stenosis. A cardiac CTA was then performed, which showed the large RCA fistula communicating with the CS; the posterior interventricular artery originated from the RCA beyond the connection to the CS and was of normal calibre (*Figure 1*). The coronary veins were noted to be dilated in the whole coronary tree. The CS was significantly dilated (up to 39 by 48 mm in diameter), with a relatively stenotic connection to the right atrium (4.5 mm).

It was proposed that the stenosed ostium of the CS was playing a significant role in her anginal symptoms by increasing coronary venous pressure, thus decreasing coronary perfusion pressure globally, rather than symptoms from RCA ischaemia alone due to the fistula, which she had tolerated well previously. Medical therapy was initiated with a calcium channel blocker, with only slight improvement in symptoms. An operation for ligation of the fistula, bypass graft to the posterior interventricular artery and repair of the CS ostium was discussed. After multidisciplinary review, a transcatheter approach to treat the CS ostial stenosis was chosen as a first step, given her deconditioned state and her preference for a less invasive option.

Under general anaesthesia and with transesophageal echocardiographic (TEE) guidance, access was obtained in the right femoral vein and left femoral artery. She had normal pulmonary pressures, with a mean pulmonary arterial pressure of 22 mmHg and pulmonary capillary wedge pressure of 14 mmHg. There was a left-to-right shunt, with pulmonary to systemic flow ratio calculated at 1.3.

Angiography of the RCA to CS fistula showed no obvious venous effluence from the CS. The CS ostium was engaged under TEE guidance (*Figure 2*), using a curved 8F sheath and an exchange-length Glidewire through a 4F angled catheter. The catheter was advanced in the CS, and the wire exchanged for an extra-stiff wire, over which a Multitrak catheter was advanced. The initial CS pressure was 58/36, mean 40 mmHg (*Figure 3A*); at that time, aortic pressure was 104/50. The coronary ostium was dilated serially with an 8 mm × 20 Mustang balloon and then 12 mm × 20 mm and 14 mm × 20 mm Atlas non-compliant balloons, with manual inflation. The final pressure in the CS was 11 mmHg (*Figure 3B*), with no gradient to the right atrial pressure. Angiography in the RCA showed faster transient of contrast to the CS and right atrium.

The postoperative course was uneventful, and the patient reported relief from angina and was able to ambulate around the ward without symptoms the day after the procedure. At 3 months of follow-up, she remained symptom free. Transthoracic echocardiogram showed a stable CS to RA gradient.

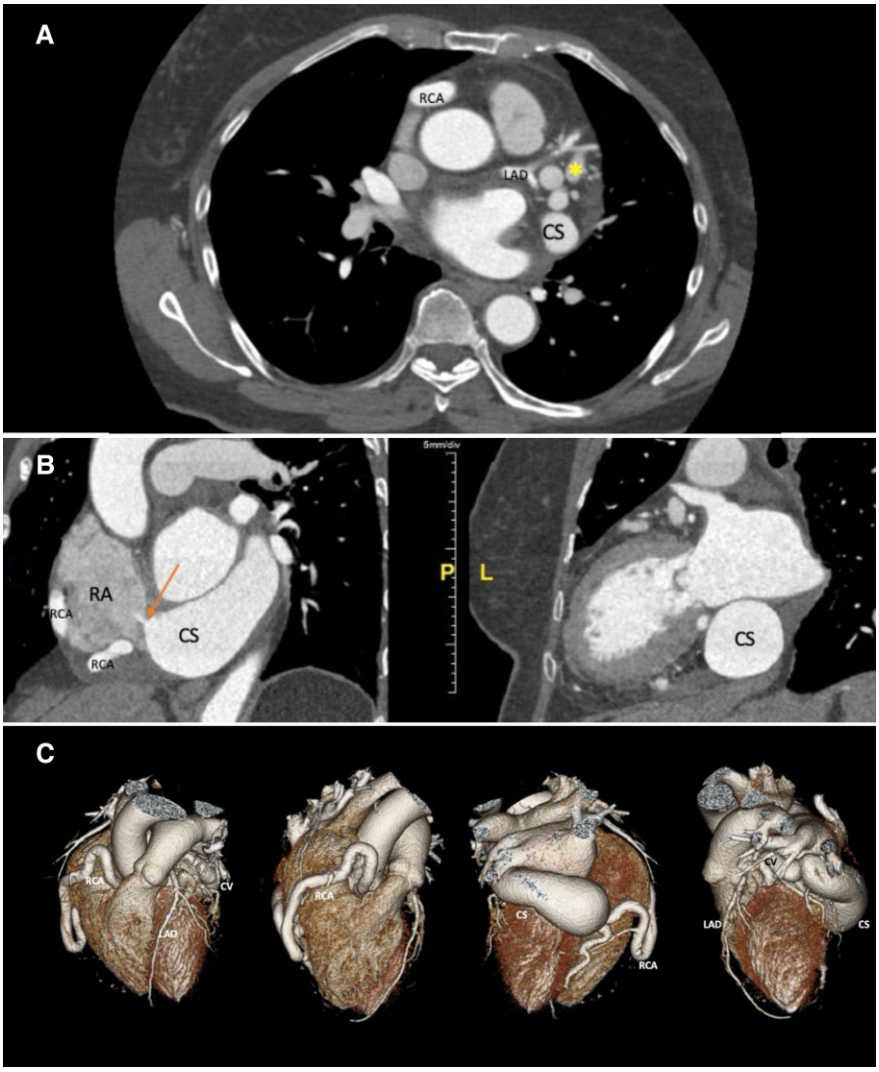


Figure 1 Cardiac computed tomography demonstrating coronary fistula, dilated coronary veins and dilated coronary sinus. Cardiac computed tomography, with representative axial (*A*), modified coronal and saggital (*B*) and three-dimensional reconstruction (*C*), demonstrating a right coronary artery to coronary sinus fistula. Dilated coronary veins are seen in (*A*), labelled with asterisk, and (*C*). CS, coronary sinus; CV, coronary vein; LAD, left anterior descending coronary artery; RCA, right coronary artery.

Discussion

Coronary fistula draining to the CS are rare[5,6] (7% of the overall 0.002% incidence of coronary fistula in the general population[1]), and association with CS ostial stenosis has been reported very infrequently. CS ostial stenosis can cause elevated coronary venous pressure, leading to decreased global coronary perfusion and symptoms of angina or heart failure. Previous case reports of coronary fistula and CA ostial stenosis were treated with either medical therapy or surgery,[7,8] and our case is the first to our knowledge to report successful percutaneous treatment. In this case, we hypothesize the patient had global coronary venous hypertension (30 mmHg in diastole) leading to a reduction in coronary perfusion pressure (to 20 mmHg as her systemic diastolic pressure was 50 mmHg, as opposed to 40–60 mmHg normally). In contrast, CS flow reducer devices have been used for treatment of refractory angina due to coronary arterial stenosis; it is thought the increase in coronary venous pressure following device implantation leads to a decrease in perfusion in 'healthy' coronary segments and relative increase in perfusion in the territory of the diseased coronary, thus leading to decrease in angina.[9]

Balloon dilation of CS stenosis has been reported to be safely performed in some patients with stenosis following lead placement for cardiac resynchronization therapy[10] or epicardial ablation.[11] Our patient had had an ablation for an atrioventricular nodal re-entrant tachycardia a few years prior to presentation; a dilated CS was already noted on CTA prior to that procedure. It is possible however that the endocardial ablation may have contributed to a fibrosing reaction at the CS ostium. After balloon angioplasty, the risk of re-narrowing and recurrent stenosis is not well known and is likely to

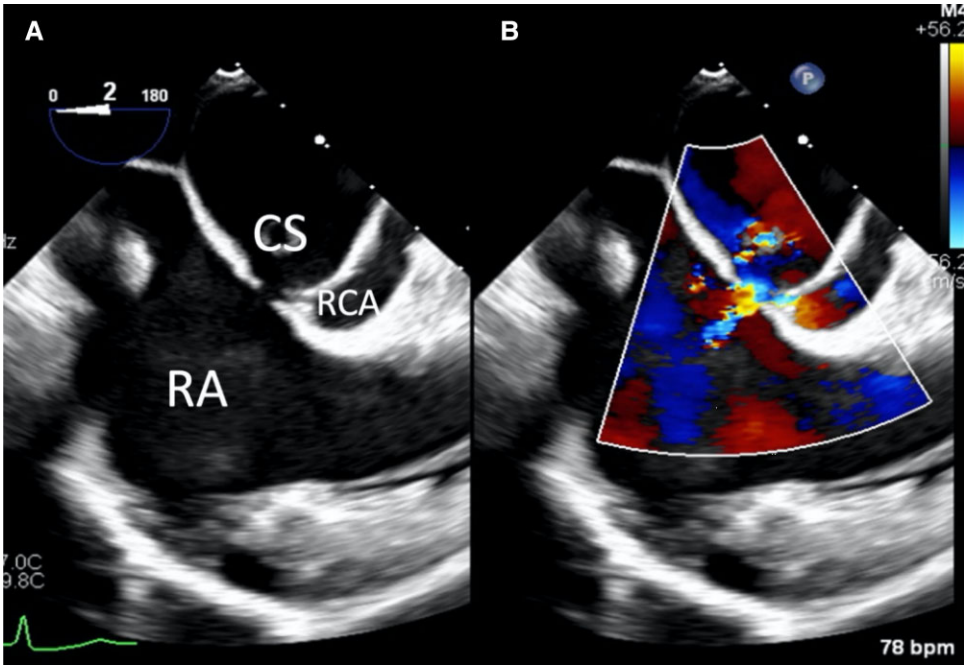


Figure 2 Transesophageal echocardiogram. Intraprocedural transesophageal echocardiogram, in two-dimensional (*A*) and color Doppler (*B*), showing the stenosed ostium of the coronary sinus with a high velocity efflux from the coronary sinus to the right atrium. A portion of the dilated right coronary artery is seen near the coronary sinus. CS, coronary sinus; RA, right atrium; RCA, right coronary artery.

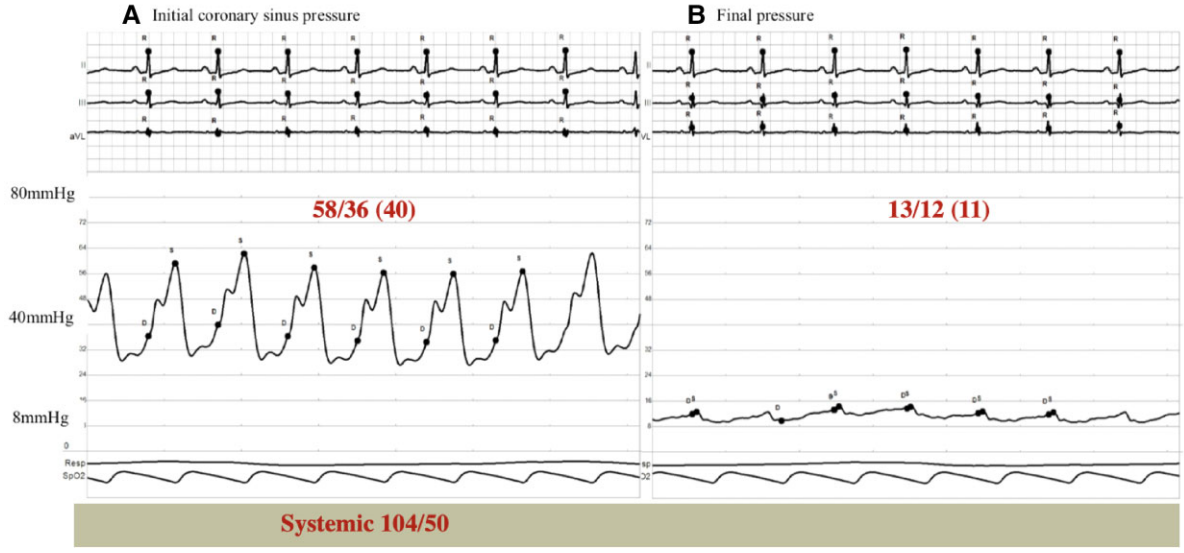


Figure 3 Intraprocedural haemodynamics. Initial (*A*) and final (*B*) systolic, diastolic and mean pressure in the coronary sinus during the procedure, with systemic arterial pressure as a reference (which remained stable during the procedure).

depend on the initial injury and risk of new or recurrent fibrosis. In the case of coronary fistula to the CS, we hypothesize that the high flow across the CS ostium is likely to be protective; our patient will be monitored with history, clinical exam and TTE which can visualize gradient across the CS ostium. We expect some increase in the degree of left-to-right shunting through the fistula after relief of the CS stenosis and will follow her clinically for signs of RV volume overload.

Importantly, CS ostial stenosis or atresia can be associated with other congenital defects.[12] In particular, children with CS atresia often have a left-sided superior vena cava which acts as a decompressing vein of the coronary venous system; acute ventricular dysfunction due to myocardial ischaemia has been described after left-sided superior vena cava ligation in those patients with unrecognized CS atresia.[13]

Surgical options for management of coronary fistula include ligation and consideration of bypass to distal arteries. After either surgical ligation or transcatheter device closure of the fistula, the coronary segment proximal to the fistula usually remains aneurysmal and at risk for thrombosis and myocardial infarction. Coronary bypass grafting of the distal vessels may be performed, though if there is still antegrade flow without significant stenosis, patency is lower and may not protect against long-term thrombosis.

In conclusion, we present a case of refractory angina caused by coronary venous hypertension due to CS dilation in the setting of CS ostium stenosis and an RCA to CS fistula. Our case is unique in that a percutaneous approach was successfully performed. Cross-sectional imaging prior to the procedure, intraprocedural TEE and a favourable anatomy were essential in guiding the procedure and successfully engaging the CS ostium.

Lead author biography

Dr Stefanescu Schmidt is an Adult Congenital Heart Disease specialist and Interventional Cardiologist. Her clinical practice and research focus on interventions to increase quality of life and longevity of adults with congenital heart disease.

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Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with required guidance.

Conflict of interest: E.M.H. is a consultant for Edwards, Medtronic and Abbott. He has received research grants from Abbott. The Structural Heart Disease programme at University Health Network receives educational support and fellowship funding from Abbott, Edwards and Medtronic and is a participant is clinical trials for devices from Abbott, Edwards and Medtronic.

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