ESC European Society of Cardiology
European Heart Journal - Case Reports
doi:10.1093/ehjcr/ytab121

CASE REPORT
Congenital heart disease

Successful coil embolization of a large right coronary artery-coronary sinus fistula causing a significant left-to-right shunt: a case report

Lamees I. El Nihum [1], Akanksha N. Thakkar[2], Ponraj Chinnadurai [3], and C. Huie Lin [2]*

[1]Texas A&M College of Medicine, Bryan, TX, USA; [2]DeBakey Heart & Vascular Center, Houston Methodist Hospital, Houston, TX, USA; [3]Siemens Medical Solutions USA Inc., Hoffman Estates, IL, USA

For the podcast associated with this article, please visit https://academic.oup.com/ehjcr/pages/podcast

Received 4 November 2020; first decision 25 November 2020; accepted 12 March 2021

Background This case reviews a challenging but successful transcatheter coil embolization of a large congenital coronary artery fistula (CAF) causing a significant left-to-right shunt.

Case summary A 51-year-old female with no significant prior history presented with symptoms of dyspnoea and chest discomfort. Extensive evaluation revealed a large CAF between a tortuous right coronary artery (RCA) and the coronary sinus (CS) composed of three aneurysmal pseudochambers. Closure of the RCA-CS fistula was attempted through coil deployment into the fistula neck. However, due to the brisk flow through the fistula, both coils embolized into the fistula sac. An alternative location was subsequently identified on three-dimensional rendering of a computed tomography angiography scan, which revealed a sharp bend in the RCA prior to the fistula neck and distal to the posterior descending artery (PDA) takeoff. Repeat attempt at embolization was accomplished using a telescoping system to reach and occlude the targeted bend. The coil mass remained stable and angiography demonstrated reduced flow through the fistula and preserved patency of the PDA. The decreased residual flow through the fistula secondary to the initial embolization attempt likely aided the successful deployment of coils in the second and final attempt. At 1 year, the patient was doing well with resolution of her symptoms and no clinical symptoms of coronary ischaemia.

Discussion We suggest that an initial unsuccessful attempt at transcatheter embolization of a CAF should not preclude subsequent attempts for closure when there exists an appropriate indication.

Keywords Embolization • Therapeutic • Coronary vessels • Heart defects • Congenital • Heart aneurysm • Heart catheterization • Case report

* Corresponding author. Tel: 713-441-4333, Fax: 713-790-3023, Email: clin@houstonmethodist.org
Handling Editor: Mohammed Al-Hijji
Peer-reviewers: Elad Asher and Flemming Javier Olsen
Compliance Editor: Carlos Minguito Carazo
Supplementary Material Editor: Fabienne Vervaat

Learning points

- Coronary artery fistulas (CAFs) are rare congenital anomalies for which the ideal management is not well studied.
- Large CAFs may present with multiple high-flow chambers that complicate attempts at transcatheter embolization. However, reduction in residual flow through the fistula secondary to initial coil embolization may aid subsequent attempts.
- An initial unsuccessful attempt at transcatheter embolization should not preclude subsequent attempts for fistula closure.

Introduction

Coronary artery fistulas (CAFs) are rare congenital anomalies; incidence varies from 0.002% to 0.3%.[1] The right coronary artery (RCA) is reportedly the most common site of origin of CAFs, accounting for 50–55% of cases; fistulas draining to the coronary sinus (CS) account for 7% of cases.[1,2] Though most CAFs are asymptomatic, symptoms of CAFs include dyspnoea on exertion and palpitations.[1] The ideal management of CAFs is not well studied due to the rarity of the condition. Previously, the presence of a large CAF was a class I indication for surgical or transcatheter closure even in the absence of symptoms.[2] However, observational data have shown that closure may have significant implications and must be considered on a case-by-case basis.[3,4] We present the case of a patient with a large RCA-CS fistula who underwent an initial unsuccessful transcatheter coil embolization attempt that helped guide a second, successful transcatheter embolization of the fistula.

Timeline

Presentation	Dyspnoea, chest discomfort, and atrial flutter
One month later	Transthoracic echocardiogram: right atrial and right ventricular enlargement
	Catheterization: Q_P:Q_S 1.4
	Computed tomography angiography (CTA): large, tortuous fistula between the right coronary artery (RCA) and the coronary sinus (CS) composed of three aneurysmal pseudochambers
Three months	Initial attempt to embolize RCA-CS fistula at the fistula neck
Four months	Atrial flutter ablation with no subsequent episodes of palpitations
Six months	Continued dyspnoea
Eight months	Transthoracic echocardiogram: coils from initial embolization procedure lodged within fistula sac
	Second and final coil embolization of RCA-CS fistula through an alternate location identified on three-dimensional CTA with preservation of the posterior descending artery
Twenty months	Clinically well and asymptomatic with no symptoms of coronary ischaemia
	Cardiac magnetic resonance imaging: Q_P:Q_S 1.1

Case presentation

A 51-year-old female with no significant prior history presented as an outpatient with dyspnoea on exertion and palpitations secondary to atrial flutter. Physical exam revealed a systolic murmur with a grade of 2/6. Transthoracic echocardiogram revealed moderate right atrial (RA) and right ventricular enlargement with normal systolic function. Right heart catheterization revealed a left-to-right shunt with step-up in oxygen saturation from the superior vena cava (SVC) (73%) to the pulmonary artery (80%). Q_P:Q_S was 1.4 and pulmonary vascular resistance was 1.4 WU. Selective coronary angiogram revealed a tortuous, ectactic RCA with an aneurysmal connection through three pseudochambers to the CS and subsequent drainage into the RA (*Figure 1*). The posterior descending artery (PDA) takeoff appeared normal (*Figure 1*). Venography revealed a severely dilated SVC. Computed tomography confirmed the presence of three pseudochambers constituting the fistula sac: a 5 cm chamber immediately distal to the RCA, followed by a 2nd and 3rd chamber connected by a narrow neck (*Figures 2* and *3*).

Given the significant left-to-right shunt causing right heart enlargement, which was the likely aetiology for her atrial flutter, a decision was made to treat the patient's fistula. Multidisciplinary discussion with cardiac surgeons led to the decision of a transcatheter approach given the extensive reports of coronary thrombosis following surgery in this particular scenario. The patient was taken to the catheterization lab and coiling of the RCA-CS fistula neck was attempted by deploying a 12 mm × 60 cm packing coil and a 14 mm × 60 cm packing coil (Penumbra, Inc., CA, USA). However, due to the brisk flow through the fistula, both coils embolized into the fistula sac (*Figure 4A*).

The patient later underwent a 2nd embolization attempt. To prevent its inadvertent embolization, the PDA was identified on computed tomography angiography (CTA) pre-procedure and again on fluoroscopy intra-procedurally. Importantly, the flow through the RCA seemed to have slowed with better contrast opacification during the 2nd catheterization, likely related to partial occlusion of the distal segments of the fistula. In order to overcome the tortuosity of the RCA, a telescoping delivery system was created using a 7-Fr 90 cm Flexor® guiding sheath (Cook Medical LLC, IN, USA) (*Figure 4A*) within which was advanced a Benchmark™ mother catheter (Penumbra, Inc., CA, USA) to the mid-RCA (*Figure 4B*), and a daughter catheter to the distal RCA (*Figure 4C*). A 150 cm LANTERN microcatheter (Penumbra, Inc., CA, USA) was then advanced into the fistula neck (*Figure 4D*). A stable coil mass was again attempted with deployment of a 14 mm × 34 cm AZUR framing coil (Terumo, NJ, USA) into the fistula neck (*Figure 4E*); however, this also embolized into the fistula sac (*Figure 4F*).

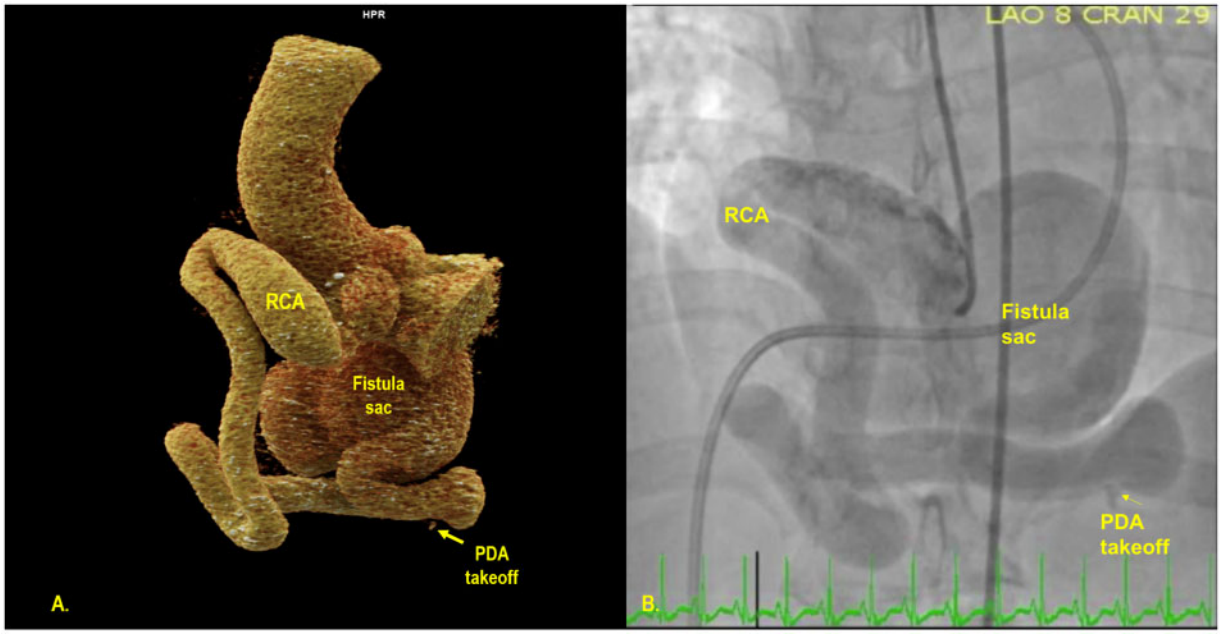

Figure 1 Three-dimensional cinematic rendering of computed tomography angiography (*A*) and angiography (*B*) demonstrated a tortuous, ectatic right coronary artery (RCA) with takeoff of the posterior descending artery (PDA) prior to connecting to a large fistula sac.

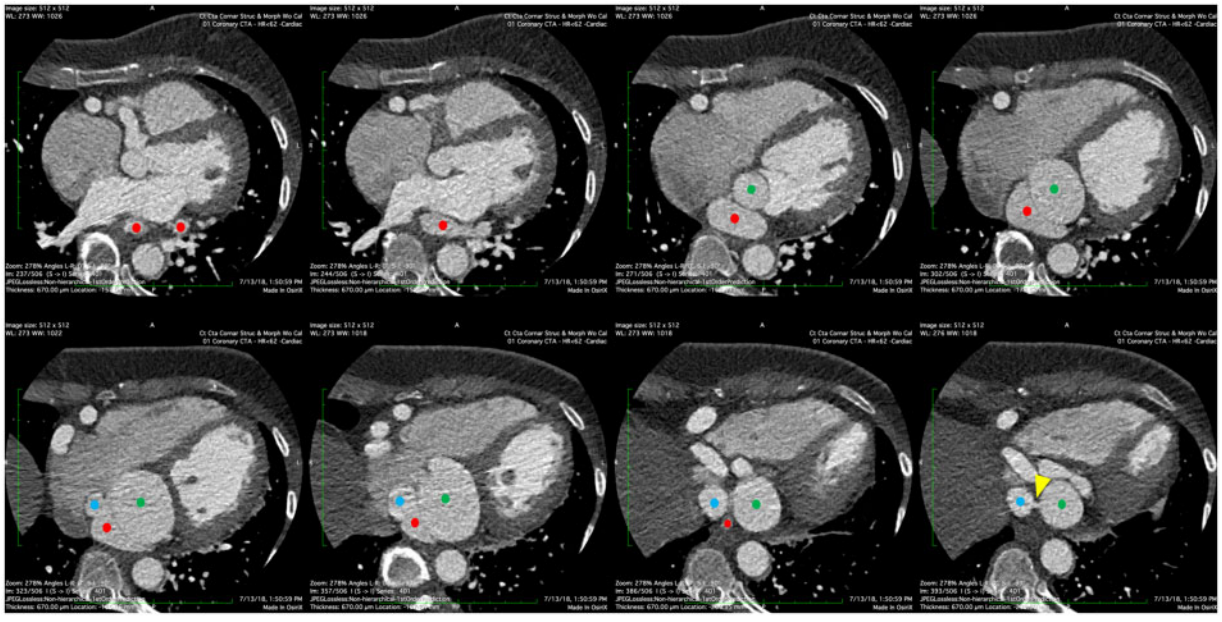

Figure 2 Craniocaudal computed tomography sequence demonstrating 1st pseudochamber (red), 2nd pseudochamber (green), and 3rd pseudochamber (blue) of the fistula. A narrow neck connected the 2nd and 3rd pseudochambers (arrowhead).

An alternative target location for the coils was subsequently identified in a sharp bend in the RCA distal to the PDA takeoff and coursing towards the fistula sac (*Figure 4G*), into which a series of coils were rapidly deployed (*Figure 4H*). The coil mass remained stable, and angiography demonstrated reduced flow through the fistula and preserved patency of the PDA. One year later, the patient was doing well with resolution of her symptoms, no clinical symptoms of coronary ischaemia and a reduced Q_p:Q_s of 1.1.

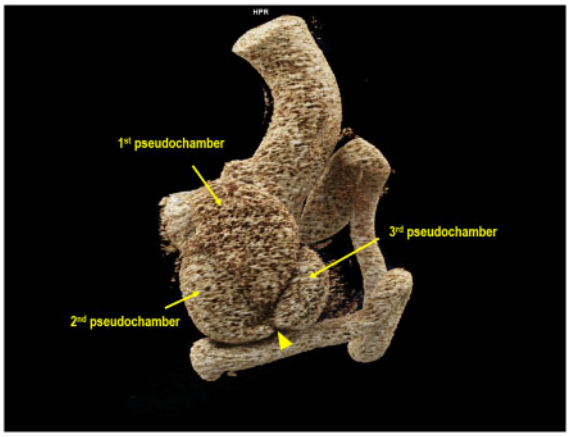

Figure 3 Three-dimensional cinematic rendering of computed tomography angiography demonstrating a narrow neck between the 2nd and 3rd pseudochambers of the fistula (arrowhead).

Discussion

Transcatheter approaches to coronary fistulas offer a safe and effective alternative to high-risk open surgical procedures. Though successfully conducted, Harris *et al.*[5] noted the challenge posed by large, tortuous, high-flow fistulas in transcatheter coil embolization repair. Indeed, a giant 4 cm × 7 cm RCA aneurysm described by Komoda *et al.*,[6] with a fistula into the CS, was surgically repaired. Due to the lack of an optimal surgical option for our patient, a transcatheter solution was offered, allowing for successful coil embolization of her large RCA-CS fistula.

We encountered multiple challenges during fistula closure. First, the tortuosity of the RCA made it difficult to advance a stable device delivery system to the fistula. Second, the fistula would require closing in a manner that would ensure preservation of the PDA. Third, we needed to achieve a stable coil mass that would withstand the brisk flow through the fistula and adequately occlude it. To address our fistula tortuosity, a telescoping delivery system was used to advance into the fistula neck. An angiographic view illustrating the PDA

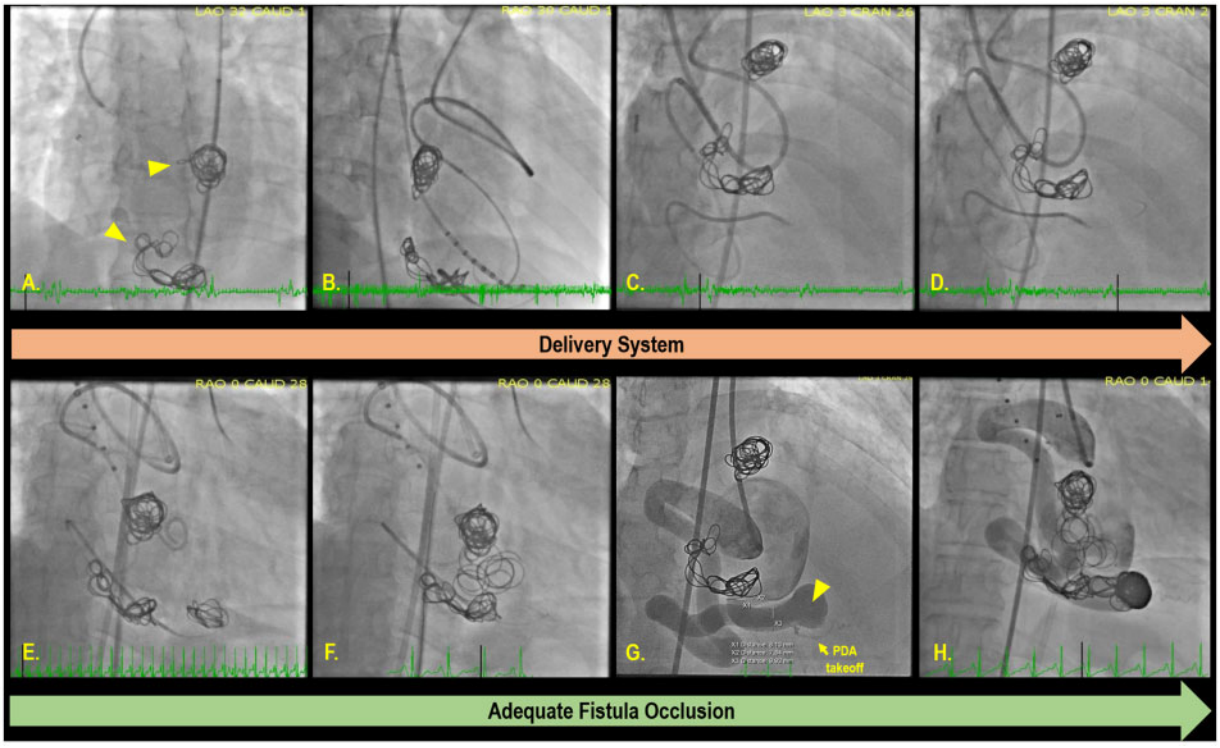

Figure 4 Angiography demonstrated coils (arrowheads) from first embolization attempt (*A*). A telescoping delivery system was created on 2nd catheterization attempt using a 7 Fr 90 cm Flexor® guiding sheath (Cook Medical LLC, IN, USA) (*A*) within which a Benchmark™ mother catheter (Penumbra, Inc., CA, USA) was advanced to the mid-right coronary artery (*B*), a daughter catheter to the distal right coronary artery (*C*) and a 150 cm LANTERN microcatheter (Penumbra, Inc., CA, USA) into the fistula neck (*D*). A 14 mm × 34 cm AZUR framing coil (Terumo, NJ, USA) was deployed (*E*) and subsequently embolized into the fistula (*F*). A target location was then identified (arrowhead) distal to the posterior descending artery (PDA) takeoff (*G*). A 16 mm × 60 cm Ruby Standard coil (Penumbra, Inc., CA, USA) was deployed, followed rapidly by a 2nd 16 mm × 60 cm Ruby Standard coil, a 14 mm × 34 cm AZUR framing coil, a 16 mm × 50 cm Ruby Soft coil, an 8 mm × 28 cm AZUR CX coil, a Penumbra packing coil, and an 8 mm × 20 cm AZUR hydrogel coil (*H*).

takeoff allowed for avoidance of inadvertent occlusion. Finally, a target location for embolization was identified on three-dimensional CTA imaging. Rather than deploying into the fistula neck, coil embolization was successfully redirected to a sharp bend in the RCA distal to the PDA takeoff and leading towards the fistula sac. A similar case was reported by Tacoy *et al.*[7] in a transcatheter embolization of a rare congenital fistula draining from the left circumflex artery (LCA) into the CS. Embolization of the severely tortuous, high-flow LCA-CS fistula was achieved through deployment of four 3 mm Guglielmi detachable coils (Boston Scientific/Target, Cork, Ireland) while maintaining patency of the LCA's distal branches.[7]

Reported long-term outcomes of CAF repair include morbidities from angina to myocardial infarction.[3] One investigation found that the sole angiographic feature predictive of adverse outcome of CAF closure was termination of the CAF into the CS.[2,3] Of the 76 patients studied, 11 underwent major complications that included coronary thrombosis; 10 of these patients had a CAF that drained into the CS, and 5 were taking antithrombotic medication at the time of diagnosis.[3] Several potential determinants of thrombosis following CAF repair have been theorized, including fistula length and diameter, coronary tortuosity and abrupt changes in vessel calibre, and abrupt cessation of high-volume flow following repair.[3] Long-term follow-up and anticoagulation should thus be initiated in patients following CAF repair.[2–4] Our patient was discharged on an anticoagulation regimen of aspirin, clopidogrel, and rivaroxaban.

Despite the challenges and complexity involved in transcatheter approaches to coronary fistulas, we propose that a transcatheter approach can be safely attempted when the procedure is indicated. Successful closure of our patient's large, high-flow fistula was potentially aided by the patient's initial embolization attempt, after which the coils had embolized into the fistula sac and caused a reduction in flow rate. Right coronary angiography performed prior to the repeat embolization revealed that the first coil sat in the 1st chamber, while the 2nd coil sat in the narrow neck between the 2nd and 3rd chambers (*Figures 2–4A*). The reduced residual flow through the fistula allowed for rapid deployment of a series of coils into a sharp bend in the RCA distal to the PDA takeoff, thus reducing flow through the fistula and preserving flow to the PDA. We therefore suggest that an initial unsuccessful attempt at transcatheter embolization of a CAF should not preclude subsequent attempts for closure when there exists an appropriate indication.

Lead author biography

Lamees I. El Nihum is a medical student at the Texas A&M University College of Medicine in Texas, USA, and a Pilot Student in the college's EnMed (Engineering & Medicine) Programme. She completed a BS in chemical engineering at Texas A&M University with a minor in aerospace engineering, presenting a thesis on "Vertical-lift aircraft: An application of computational fluid dynamics to a VTOL engine model" (2015) as an Undergraduate Research Scholar. She is currently conducting research at the Houston Methodist DeBakey Heart & Vascular Center as a Burroughs Wellcome Fund scholar in the Texas A&M University Academy of Physician Scientists, where she is studying diagnostic applications of computational fluid dynamics in congenital heart disease.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Acknowledgements

The authors would like to thank Amish S. Dave, MD: Houston Methodist DeBakey Cardiology Associates, Houston, TX, USA; Gary A. Monteiro, MD: Houston Methodist Hospital, Houston, TX, USA; and John P. Breinholt, MD; Phoenix Children's Hospital, Phoenix, AZ, USA, for their contributions to this article.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: C.H.L., MD, PhD, is a data monitoring committee member of ACI Clinical, speaker for Abiomed, proctor for Abbott, and course director for Gore Medical. P.C. is a full-time Research Collaborations Manager and Senior Key Expert at Advanced Therapies Division, Siemens Medical Solutions USA, Inc.

Funding: L.I.E. was supported by a Burroughs Wellcome Fund Physician Scientist Award to the Texas A&M University Academy of Physician Scientists.

References

1. Romano S, Petroni R. Coronary artery fistula: an innocent bystander or harmful company? *Kardiol Pol* 2019;**77**:1009–1010.
2. Yun G, Nam TH, Chun EJ. Coronary artery fistulas: pathophysiology, imaging findings, and management. *Radiographics* 2018;**38**:688–703.
3. Valente AM, Lock JE, Gauvreau K, Rodriguez-Huertas E, Joyce C, Armsby L et al. Predictors of long-term adverse outcomes in patients with congenital coronary artery fistulae. *Circ Cardiovasc Interv* 2010;**3**:134–139.
4. Said SM, Burkhart HM, Schaff HV, Connolly HM, Phillips SD, Suri RM et al. Late outcome of repair of congenital coronary artery fistulas—a word of caution. *J Thorac Cardiovasc Surg* 2013;**145**:455–460.
5. Harris WO, Andrews JC, Nichols DA, Holmes DR. Percutaneous transcatheter embolization of coronary arteriovenous fistulas. *Mayo Clin Proc* 1996;**71**:37–42.
6. Komoda S, Komoda T, Ivanitskaia-Kuehn E, Dreysse S, Pasic M, Hetzer R. Giant aneurysm of the right coronary artery and fistula to the coronary sinus. *Gen Thorac Cardiovasc Surg* 2010;**58**:78–81.
7. Tacoy G, Ebinc H, Onal B, Abaci A, Ilgit E, Yalçin R. Congenitally severe tortuous circumflex artery fistula draining into the coronary sinus: transcatheter closure with Guglielmi detachable coils via different delivery system. *J Cardiol* 2009;**54**:317–321.