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Percutaneous closure of giant aneurysmal coronary artery-to-coronary sinus fistulae with guidance from three-dimensional printed models: a case series

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Background Coronary artery fistulae are abnormal communications of coronary arteries with systemic vasculature, pulmonary vasculature, or cardiac chambers. Use of multimodality imaging can be paramount to understanding anatomical and functional features of these complex vascular lesions, therefore optimizing success of potential curative interventions.

Case summary We present two patients with incidentally discovered giant aneurysmal coronary arteries with distal fistulous connections to the coronary sinus, which were successfully closed percutaneously with Amplatzer Septal Occluders using the assistance of three-dimensional (3D) printed heart models.

Conclusion Computed tomography–guided reconstruction with 3D multiplanar, multicolour printed models can help augment visuospatial understanding of the size, origin, course, and drainage of giant aneurysmal coronary artery-to-coronary sinus fistulae, and with manual bench testing can assist with choosing accurately sized and shaped devices for closure.

Keywords Coronary fistula • Giant coronary aneurysm • Computed tomography • Three-dimensional printed model • Case series

ESC Curriculum 3.4 Coronary angiography • 2.4 Cardiac computed tomography • 9.7 Adult congenital heart disease

Learning points

- Giant aneurysmal coronary arteries with fistulous connections to the coronary sinus can lead to myocardial ischaemia and heart failure due to coronary steal and left-to-right shunting.
- Computed tomography–guided reconstruction with 3D multiplanar printed models can help augment visuospatial understanding of the size, origin, course, and drainage of these fistulae. Manual bench testing of different sized and shaped occluder devices with printed models can confirm adequate seal and rims for closure.

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Introduction

Coronary artery fistulae (CAF) are abnormal vascular communications of coronary arteries without associated capillary beds. Incidence is ~0.9% with the majority being congenital or sporadic, and 50–55% originate from the right coronary artery (RCA). If a CAF originates within the proximal one-third of the artery, typically only the proximal feeding arteries tend to be dilated. Distal CAFs tend to have dilation of the entire vessel leading to aneurysm formation.[1]

CAFs are classified into coronary cameral fistulae that drain into lower pressure cardiac chambers, or coronary arteriovenous fistulae that drain into segments of the pulmonary or systemic circulation. Coronary artery-to-coronary sinus (CS) fistulae are one type of coronary arteriovenous fistula, and are the third most common type of CAF. These patients can also present with dilated aneurysmal CSs.[1]

Use of multimodality imaging can be paramount to understanding anatomical and functional features of these complex vascular lesions, therefore optimizing success of potential curative interventions.[2] We present two patients with giant aneurysmal coronary arteries with distal fistulous connections to the CS, which were closed percutaneously with Amplatzer Septal Occluders using the assistance of three-dimensional (3D) printed models.

Timeline

Case 1	
Week 0	Incidental discovery of coronary aneurysm on computed tomography (CT) abdomen/pelvis.
Weeks 5–8	Coronary CT angiography (CTA) revealed giant aneurysmal RCA and right posterior lateral (RPL) with fistulous connection to severely dilated CS. Echo showed severely dilated CS, low normal LV function, and four-chamber dilatation. Myocardial perfusion showed mild reversible defects of mid-distal inferior and inferolateral walls.
Week 10	Diagnostic coronary angiography confirmed CTA findings.
Week 12	Bench testing with 3D printed models supported use of 8–10 mm Amplatzer Septal Occluder.
Week 15	Successful percutaneous closure of fistula with 8 mm Amplatzer Septal Occluder under angiographic and transesophageal echocardiogram (TEE) guidance.
Case 2	
Week 0	Incidental discovery of pulsatile cardiac mass during pericardial window.
Weeks 3–5	Coronary CTA revealed aneurysmal left main and giant LCx aneurysm distally with fistulous connection to severely dilated CS. Echo showed severely dilated CS with turbulent flow into the right atrium, normal LV function, normal chamber sizes, and mild pulmonary hypertension. Myocardial perfusion showed small fixed apical defect.
Week 8	Diagnostic coronary angiography confirmed CTA findings.
Week 10	Bench testing with 3D printed models supported use of 5 mm Amplatzer Septal Occluder.
Week 14	Successful percutaneous closure of fistula with 5 mm Amplatzer Septal Occluder under angiographic and TEE guidance.

Case presentations

Case 1

A 40-year-old asymptomatic, previously healthy male presented with concern for coronary aneurysm incidentally seen on CT abdomen/pelvis performed for abdominal pain. Echocardiogram showed a severely dilated CS, low normal left ventricular function, and four-chamber dilatation. Myocardial perfusion showed mild reversible defects of the mid-to-distal inferior and inferolateral walls.

Coronary CTA revealed a giant aneurysmal RCA (proximal 1.9 × 1.5 cm, mid 1.9 × 1.8 cm, distal 1.8 × 1.4 cm). The RPL branch was severely aneurysmal (4.3 × 3.8 cm) with fistulous connection to a severely dilated CS (5.9 × 5.0 cm; *Figure 1A*). There was no associated persistent left-sided superior vena cava (SVC) or CS unroofing. Multiplanar 3D printout models were generated based on CT and demonstrated the fistulous connection to be singular, circular in anatomy, and ~6 mm in diameter (*Figure 2A*). Bench testing using different closure devices was performed with favourable anatomy for an 8–10 mm Amplatzer Septal Occluder with adequate seating and rim apposition (*Figure 2B*).

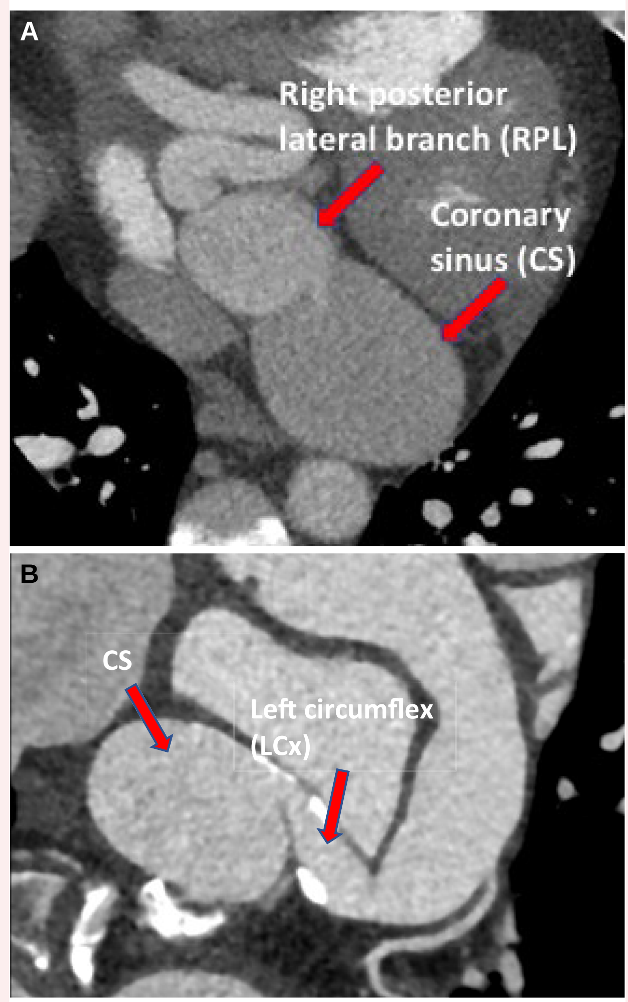

Figure 1 Coronary computed tomography angiography. (*A*) Case 1: Giant aneurysmal right coronary artery and severely aneurysmal right posterior lateral branch with fistulous connection to severely dilated coronary sinus, (*B*) Case 2: Giant aneurysmal LCx with fistulous connection to severely dilated coronary sinus.

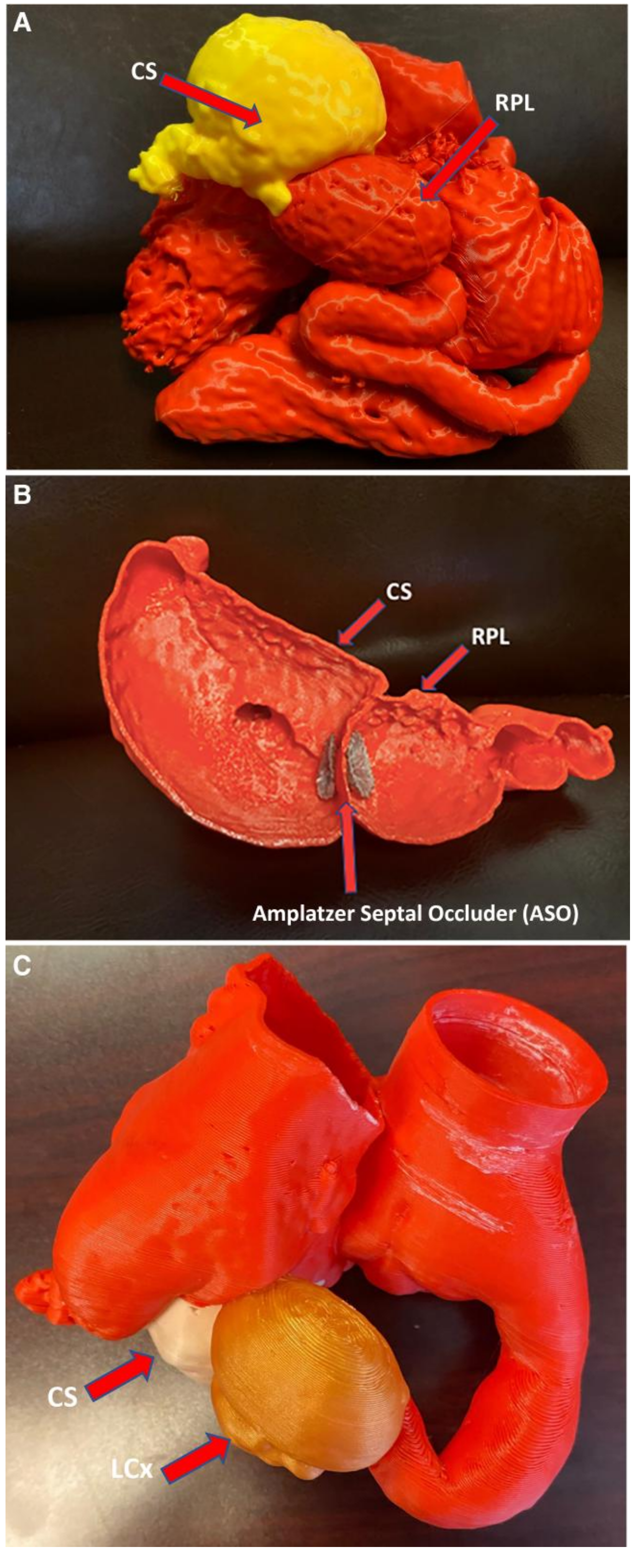

Figure 2 Three-dimensional printout models. (*A*) Case 1: Three-dimensional model demonstrating fistula to be circular and ∼6 mm in diameter. (*B*) Case 1: 8 mm Amplatzer Septal Occluder with adequate seating and rim apposition within the fistula. (*C*) Case 2: Three-dimensional model demonstrating fistula to be circular and ∼4 mm in diameter.

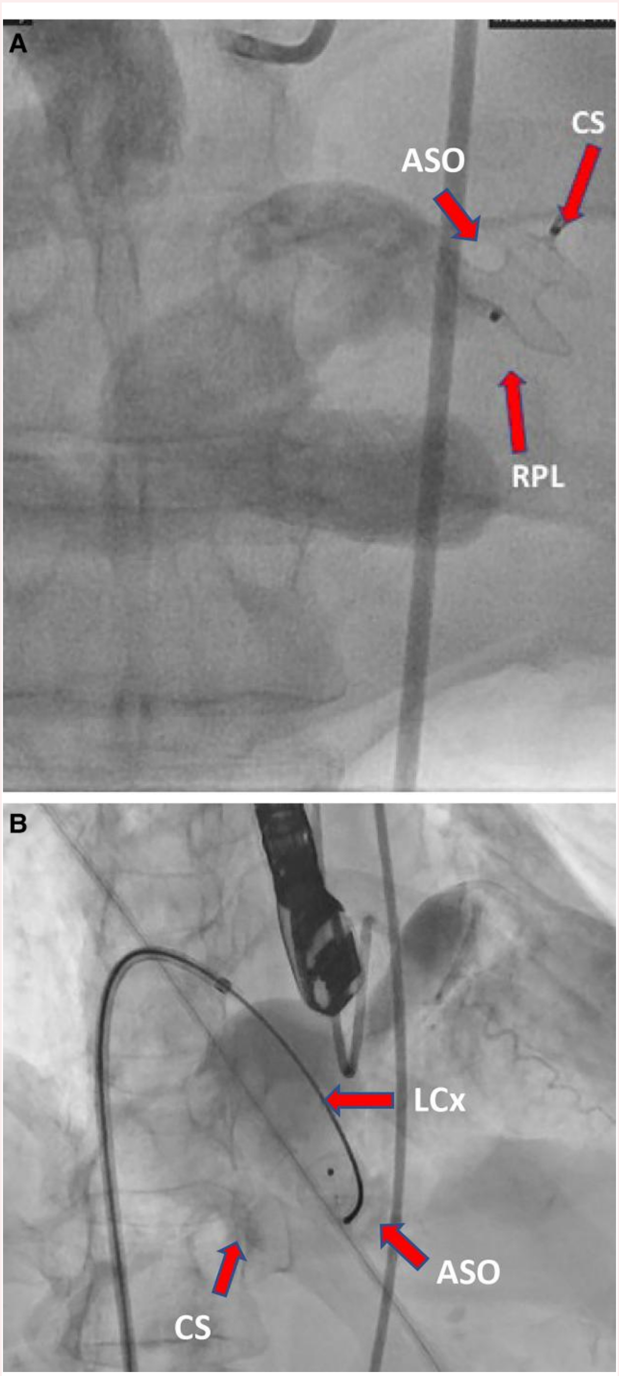

Figure 3 Percutaneous closure of fistula. Amplatzer Septal Occluder device released with significantly reduced flow into the coronary sinus. (*A*) Case 1. (*B*) Case 2.

Cardiac catheterization confirmed a giant aneurysmal RCA with fistulous connection of the RPL to a dilated CS (*Figure 3A*). Epicardial coronaries were without anomalous course or atherosclerosis. Qp:Qs ratio was 2:1 suggesting a large left-right shunt. Right heart pressures were remarkable for mild pre-capillary pulmonary hypertension [PA 41/19 (26), pulmonary capillary wedge pressure 11] in the setting of increased pulmonary blood flow. Due to large size of the fistula, significant left-right shunting with consequent chamber dilatation, and presence of coronary steal, it was decided to pursue fistula closure.

An 8 French Hockey stick guide and Caravel microcatheter were used to cannulate the RCA through which a 300 cm Runthrough wire was delivered to the RPL, and subsequently through the fistula into the CS. A 7 French Torqvue venous sheath was used to deliver a 5 French JR-4 catheter into the CS, and a 4–8 mm En-Snare was used to capture the coronary wire and exteriorize it through the venous sheath creating an arteriovenous rail. After confirming sizing with a 6×20 mm balloon, the 180° Torqvue system was used to load and deploy an 8 mm Amplatzer Septal Occluder across the fistula under angiographic and TEE guidance. The device was released without impingement on adjacent structures, and with significantly reduced flow into the CS. Final angiography showed complete closure of the fistula (*Figures 3A* and *4*). Patient was discharged on aspirin and warfarin. After 6 months patient remained asymptomatic, and repeat myocardial perfusion showed resolution of reversible perfusion defects.

Case 2

A 64-year-old female with history of hypertension, atrial fibrillation, and small cell lung cancer status post chemotherapy and radiation presented with concern for a pulsatile cardiac mass discovered incidentally during a pericardial window which was performed in the setting of exertional dyspnoea and recurrent pericardial effusions. Echocardiogram showed a severely dilated CS with turbulent flow into the right atrium, normal left ventricular function, normal chamber sizes, and mild pulmonary hypertension. Myocardial perfusion showed a small fixed apical defect.

Coronary CTA revealed an aneurysmal left main artery (1.6×1.4 cm) and a giant left circumflex artery (LCx) aneurysm distally (proximal 1.4×1.2 cm, mid 1.7×1.6 cm, distal up to 3.5 cm with inlet 0.9×0.5 cm and outlet 1.8×1.5 cm) with fistulous connection to a severely dilated CS (4.1 cm with inlet 0.3×0.3 cm and outlet 0.5×0.4 cm; *Figure 1B*). Multiplanar 3D printout models demonstrated the fistula to be singular, circular in anatomy, and ∼4 mm in diameter (*Figure 2C*). Bench testing of closure devices favoured a 5 mm Amplatzer Septal Occluder.

Figure 4 Coronary computed tomography angiography. Confirmation of Amplatzer Septal Occluder device placement in Case 1.

Cardiac catheterization revealed a giant aneurysmal LCx with fistulous connection to an aneurysmal segment of the CS. Epicardial coronaries were without anomalous course or atherosclerosis. Qp:Qs ratio was 1:1 and right heart filling pressures were normal. The decision was made to pursue intervention due to the large size of the fistula.

An 8 French EBU 3.5 guide and Caravel microcatheter were used to cannulate the LCx through which a 300 cm Runthrough wire was delivered into the fistulous connection to the CS. An 8 French femoral venous sheath was used to deliver a 5 French JR-4 catheter into the CS, and a 7 mm Micro-goose neck snare was used to capture the coronary wire and exteriorize it through the sheath creating an arteriovenous rail. After a 4 × 6 mm balloon was taken on wire to confirm sizing, the 180° Torqvue system was used to load and deploy a 5 mm Amplatzer Septal Occluder across the fistula under angiographic and TEE guidance. The device was released without impingement on adjacent structures, and final angiography showed complete closure of the fistula (*Figure 3B*). Patient was discharged on aspirin and lovenox, as well as 30 days of clopidogrel. After 4 months, patient reported no symptoms and repeat echocardiogram confirmed interval reduction in flow between the CS and right atrium.

Discussion

This case series reports the procedural success of two coronary fistula closures using Amplatzer Septal Occluders with guidance from 3D printed models. Both patients had giant aneurysmal coronary arteries with distal fistulous connections to the CS.

CAFs can present clinically as heart failure and pulmonary hypertension if there is significant left-to-right shunting, or myocardial ischaemia if the fistula impairs distal perfusion with coronary steal. Spontaneous closure of CAFs happens in 1–2% of cases but repair is warranted in those with large fistulae, presence of significant chamber dilation, or small to moderate size fistulae with evidence of myocardial ischaemia, arrhythmias, ventricular dysfunction, and/or endarteritis. Treatment can involve surgical ligation in the setting of large symptomatic fistulae with high blood flow, and those with multiple communications and drainage sites. Percutaneous transcatheter closure can be attempted if there are a finite number of drainage sites, the distal portion of the fistula is accessible with a closure device, termination of the fistula is away from normal coronary arteries, and/or if there is high surgical risk.[1] Our cases were deemed appropriate for closure in the setting of large fistula size, left-right shunting, and coronary steal, and percutaneous transcatheter methods were preferred given single drainage sites with accessible distal portions.

Aneurysmal coronary segments at distal fistulous attachments can be technically challenging for closure especially if the connection is small in diameter, often requiring creation of an arteriovenous rail to allow for venous introduction of devices. Also, fistulas terminating in the CS have anatomically driven periprocedural complications with higher rates of myocardial infarction, cardiomyopathy, and coronary thrombosis for which patients are often placed on oral anticoagulation.[1] In the setting of these difficulties, production of 3D reconstructed models can greatly enhance procedural planning with true appreciation of the complexities of these vascular anomalies. The general course and origin of arteriovenous fistulae can be delineated with CT, but angiography alone may not provide adequate visual understanding of size, number of fistulous connections, and anatomical path. Three-dimensional models allow for manual bench testing of different sized and shaped occluder devices to confirm adequate seal and stable rims for closure.[2] For our cases, 3D models allowed us to test several different devices including Amplatzer VSD occluder, septal occluder, and vascular plug-II. Based on bench testing, the morphology of the Amplatzer septal occluder was deemed most conducive to the anatomy for both cases. Pretesting allowed us to accurately preselect the devices which translated into reduced procedural times for the patients.

In addition to congenital disease, 3D reconstructed models have been used in periprocedural planning for other structural heart interventions like complex transcatheter valve cases, paravalvular leak closure and atypical left atrial appendage occlusion (LAA-O). For example, 3D printed models have been used to simulate transcatheter aortic valve replacement as well as to anticipate complications such as annular rupture, paravalvular regurgitation, need for pacemaker placement, and coronary artery occlusion. While transoesophageal echocardiography alone was reported to undersize LAA-O devices in 45% of patients, the addition of 3D printed models has improved preprocedural sizing and operator success.[3]

The 3D reconstruction involves volumetric image acquisition from modalities such as CT, data postprocessing with segmentation and modelling, selection of the type of printer and material to be used, and finally the actual process of printing.[3] Use of this technology can improve procedural efficiency and overall safety as evidenced by our two cases, but there are some limitations to acknowledge; lack of randomized data and standardized protocols/guidelines for printing, dependence on quality of original CT images for model accuracy, no currently available material to perfectly replicate the mechanical properties of human tissue, and no financing or reimbursement models to seamlessly implement this practice.[3] However, given its value in periprocedural planning, it may be worthwhile for a busy structural heart disease programme to consider investing in a 3D printing laboratory under the supervision of a dedicated multimodality cardiac imaging specialist.

Conclusion

CT-guided reconstruction with 3D multiplanar, multicolour printed models can help augment visuospatial understanding of the size, origin, course, and drainage of giant aneurysmal coronary artery-to-CS fistulae, and with manual bench testing can assist with choosing accurately sized and shaped devices for closure.

Lead author biography

Jaya Kanduri is a third-year general cardiovascular disease fellow at New York Presbyterian-Weill Cornell Medical Center in New York City, NY. She will be staying at Weill Cornell for her interventional year in 2023. She graduated from Rutgers Robert Wood Johnson Medical School in Piscataway, NJ and completed residency training at Beth Israel Deaconess Medical Center in Boston, MA.

Supplementary material

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

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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 patients in accordance with COPE guidelines.

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