European Heart Journal - Case Reports (2023) **8**, 1–6
https://doi.org/10.1093/ehjcr/ytad413

CASE SERIES
Cardiovascular imaging

Computed tomographic–based three-dimensional printing of giant coronary artery fistulas to guide surgical strategy: a case series

Mads Ørbæk Andersen [1], Morten H. Smerup[2], Kim Munk[3], Ulrik Markus Mortensen[3], Bjarne Linde Nørgaard [3], Morten Helvind[2], Henrik Ørbæk Andersen[2], and Jesper James Linde [1]*

[1]Department of Cardiology, Rigshospitalet, Copenhagen University Hospital, Blegdamsvej 9, 2100 Copenhagen, Denmark; [2]Department of Cardiothoracic Surgery, Rigshospitalet, Copenhagen University Hospital, Blegdamsvej 9, 2100 Copenhagen, Denmark; and [3]Department of Cardiology, Aarhus University Hospital, 8200 Århus, Denmark

Received 19 December 2022; revised 7 August 2023; accepted 16 August 2023; online publish-ahead-of-print 22 August 2023

Background Coronary artery fistulas (CAFs) are abnormal communications between the coronary arteries and the heart chambers, arteries, or veins, potentially leading to significant shunting, myocardial ischaemia and heart failure. Computed tomographic (CT) angiography or conventional invasive angiography is the reference standard for the diagnosis of coronary fistulas. The fistula anatomy can become very complex, which makes surgical or interventional planning challenging.

Case summary We report two cases of hugely dilated and tortuous coronary circumflex artery fistulas draining into the coronary sinus. Both patients were followed up for more than 10 years because of very complex coronary fistula anatomy and mild symptoms. From two-dimensional (2D) sliced CT images alone it, was uncertain whether surgery was feasible. However, since both patients had symptom progression (Patient 1 developed heart failure, and Patient 2 had recurrent pericardial effusions), three-dimensional (3D) heart models were printed for better understanding of the complex fistula anatomy and improved surgical planning. Both patients had successful surgery and symptomatic relief at follow-up.

Discussion The delay in surgery, until clinical deterioration, may partly be a consequence of a general reluctance in performing complex surgery in patients with CAFs. As of now, CT-based 3D printing has primarily been used in isolated cases. However, 3D printing is evolving rapidly and supplementing 2D sliced CT images with a physical 3D heart model may improve the anatomical understanding and pre-surgical planning that could lead to better surgical outcome.

Keywords 3D printing • Fistulas • Imaging • Case series

ESC curriculum 2.1 Imaging modalities • 7.5 Cardiac surgery • 9.7 Adult congenital heart disease

Learning points

- Three-dimensional printing offered a unique assessment of the tortuous and enormous labyrinth of coronary fistulas in the presented cases.
- Three-dimensional printing influences the decision to perform surgery.
- Three-dimensional printing may be of particular value in patients with coronary artery fistulas as the anatomy and thereby treatment options are often extremely difficult to assess on two-dimensional sliced computed tomographic images.

* Corresponding author. Tel: +45 35456396, Email: jesper.james.linde@regionh.dk
Handling Editor: Andreas Giannopoulos
Peer-reviewers: Sarv Priya; Andras Janosi; Johannes Haubold
Compliance Editor: Pok-Tin Tang

Introduction

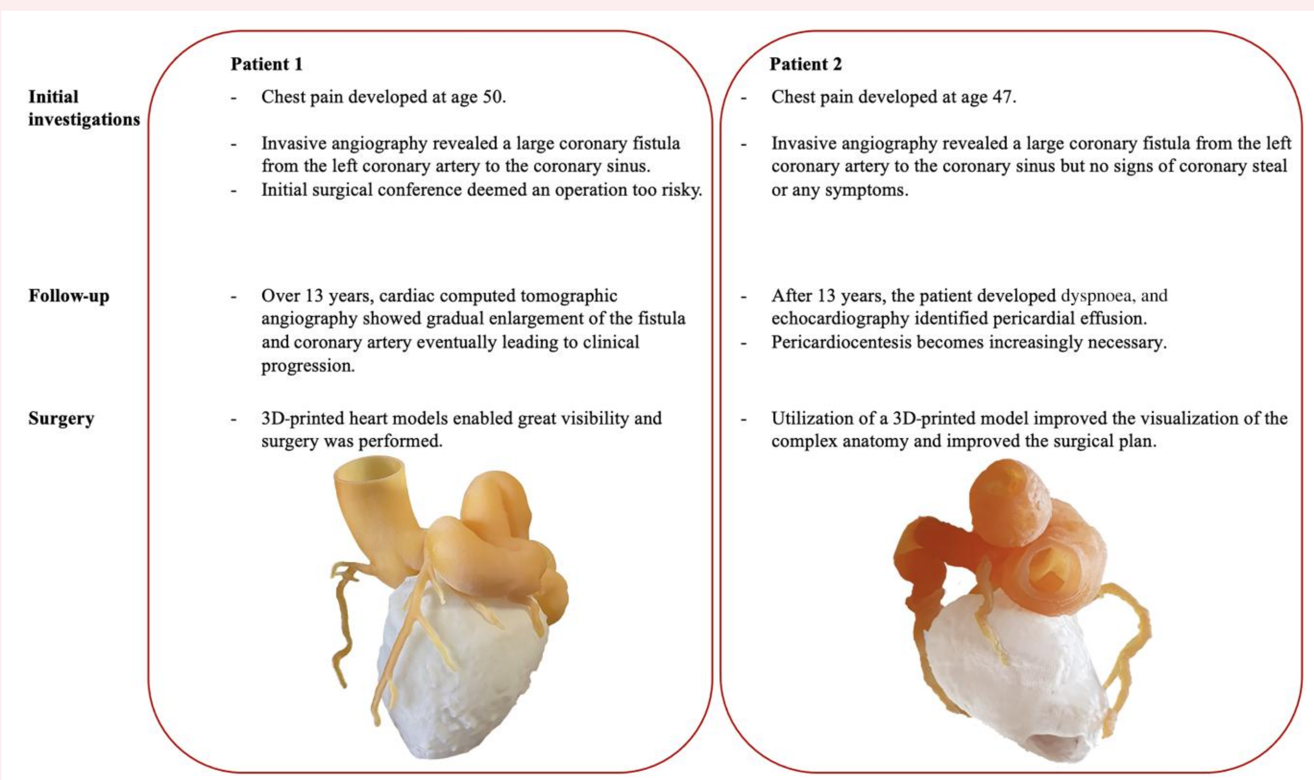
A coronary artery fistula (CAF) is defined as an abnormal communication between a coronary artery and a heart chamber, another artery, or a vein. Coronary artery fistulas can be either acquired or more often a congenital heart disease (CHD). The prevalence of CAFs has been reported to be as high as 0.9%.[1,2]

Coronary artery fistulas to the coronary sinus (CS) are the third most common type of CAF.[1,2] The progressive increase in shunt flow from the systemic arterial bed to the venous bed will often lead to a dilated and twisted course of the CAF, dilation of the CS, and unusual presentations to pericardial effusion.[1,3] Typical symptoms are dyspnoea due to shunting, left ventricular (LV) dysfunction and/or pericardial effusion, or angina pectoris caused by reduced myocardial perfusion because of a coronary steal phenomenon. When patients with CAFs become symptomatic, treatment with percutaneous trans-catheter device insertion or surgery is often recommended.[1,2] This requires an accurate demonstration of the anatomy. Computed tomographic angiography or conventional invasive angiography are reference standards for the diagnosis of CAFs. Although CT is a three-dimensional (3D) modality, the anatomy is typically displayed in two-dimensional (2D) slices.[1]

Three-dimensional printing from patient-specific CT scan has recently been introduced in the field of cardiology, especially to assess complex CHD cases.[4,5] However, 3D printing has only been reported in few patients with CAF, mainly before percutaneous treatment.[5]

We present two cases demonstrating the use of 3D printing to guide surgical planning in patients with giant CAFs.

Summary figure



Case presentation

Patient 1

Patient 1 was diagnosed with a CAF at the age of 50 after a routine X-ray revealed an enlarged heart. Invasive catheterization demonstrated a severely dilated circumflex artery (CX) fistula that drained into the CS (*Figure 1A*). The shunt ratio (*Qp*/*Qs*) was 1.4. The patient endured 200 W/8.5 METS on an exercise bicycle test and experienced no angina and had a normal clinical examination. At this point, the patient was asymptomatic except for some episodes of paroxysmal atrial fibrillation. He was followed with echocardiography and consecutive cardiac CT angiography (CCTA) over 10 years showing that the dimension of the CX fistula slowly increased (*Figure 1B–D*). Interventional or surgical closure of the CAF was discussed on multiple occasions, but due to uncertainty concerning the feasibility of surgery in combination with sparse symptoms, a conservative strategy was initially chosen with antithrombotic therapy of 75 mg aspirin daily. However, after 13 years, the patient became severely symptomatic with dyspnoea corresponding to NYHA Class III and additionally developed angina pectoris (Canadian Cardiovascular Score Class III). Angina and signs of ischaemia were judged to be results of a coronary steal phenomenon due to myocardial hypo-perfusion caused by a coronary steal phenomenon. Additionally, echocardiography now showed severe mitral regurgitation and a dilated LV. The surgical options considered were either (i) to close the fistula, including an attempt to graft branches from the fistula and perform mitral valve replacement, a with a high perioperative risk, or (ii) to list the patient for heart transplantation.

Three-dimensional print

For better understanding of the complex anatomy, the surgeons requested a CT-based 3D-printed model of the fistula and coronary

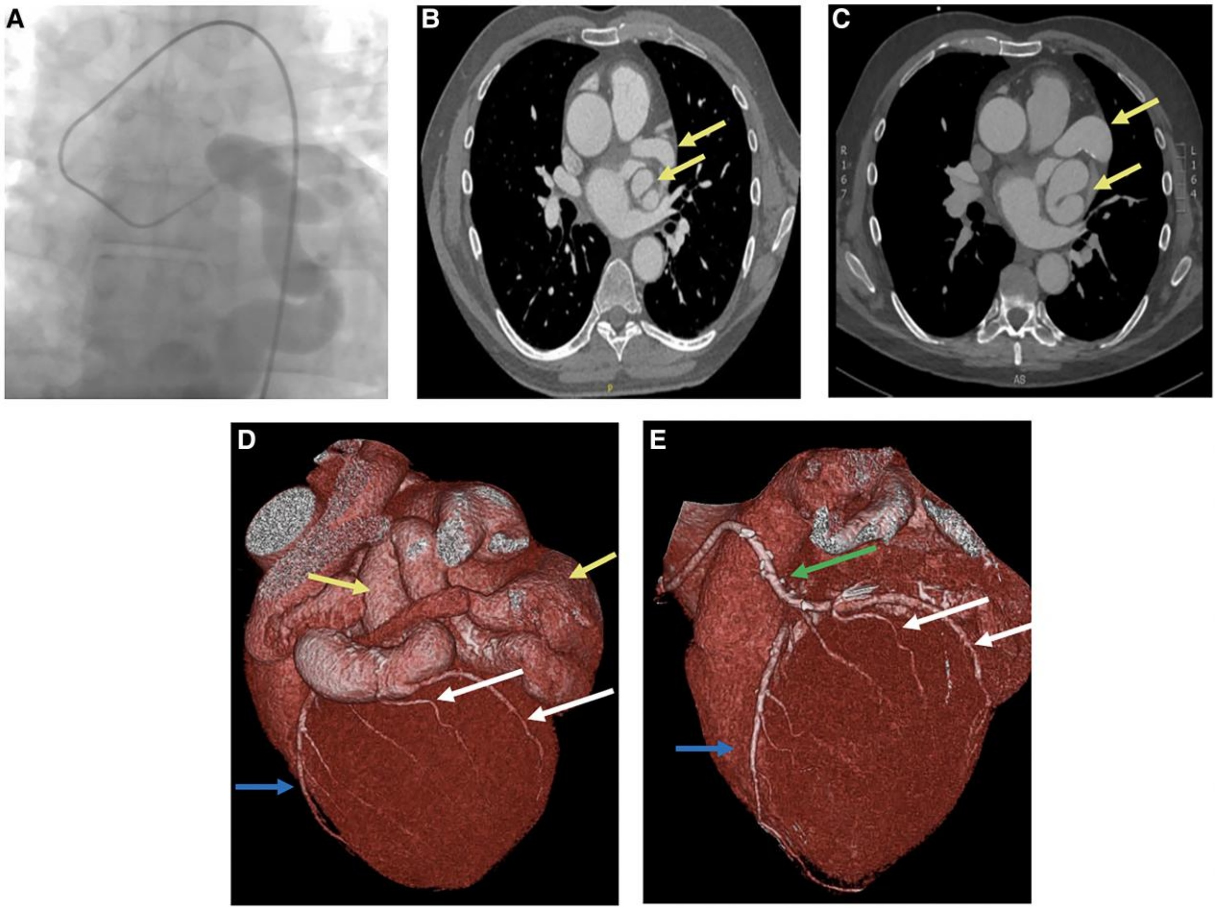


Figure 1 (*A*) Invasive catheterization demonstrated a severely dilated circumflex artery fistula. (*B–D*) Computed tomographic scans before surgery from 2009 (*B*) and 2021 (*C* and *D*) illustrating the increased dilatation of the circumflex artery fistula (yellow arrows). (*E*) Computed tomographic scan after surgery, including the venous bypass graft (green arrow) to the obtuse marginal branches (white arrows). Blue arrows points to the normal-sized left anterior descending artery.

anatomy. Computed tomographic angiography was performed using a Siemens Somatom Force scanner. A standardized electrocardiogram-triggered prospective scan protocol was used, aiming for arterial phase contrast enhancement. As per routine clinical guidelines, an oral beta-blocker was used, targeting a heart rate of <60 b.p.m. and sublingual suspension of nitroglycerin (0.8 mg) was also administered. The tube voltage was 120 kV, current was 400 mAs, and the total dose length product was 648 mGycm. Mimics Innovation Suite software (Materialise NV, Leuven, Belgium) was used to segment the model and to finally create a stereolithographic (STL) file. From the aortic root, the coronary fistula and the normal coronary arteries were segmented in a semi-automatic fashion, where the contrast-enhanced lumen was segmented in the direction going from the aortic root to the periphery and for the fistula to the connection point in the coronary sinus, which was included. After careful inspection of the segmented fistula, few additionally manual corrections were performed. Hereafter, a 'hollowing' segmentation procedure was performed, creating a fistula wall diameter of 1.5 mm. The myocardium of the left ventricle was automatically segmented, excluding the LV lumen. Segmentation of the model took approximately 2 h.

A PolyJet 3D printer (Stratasys J750) was used to print the model in an acrylic material, which took 15 h. The model included the aortic root, the coronary arteries, and CX fistula, the CS and the LV (*Figure 2*). The 3D-printed model clarified the course of the dilated, tortuous CX fistula, and its anatomic relations. From this, the surgeons judged that it would be possible to close the CX fistula proximally and to graft two major side branches from within the fistula besides performing mitral valve repair/replacement as appropriate.

Surgery

Median sternotomy was performed, and a venous graft from the left leg was harvested. Bi-caval extra-corporeal circulation (ECC) and repeated cardioplegia in the aortic root during compression of the fistula were applied. Anatomy was confirmed to be exactly as visualized on the CT scan and the 3D-printed model. After the division of the main pulmonary artery, the hugely dilated CX artery was identified and divided close to the takeoff from the left main artery (LM). Both ends were sutured so that the LM now continued directly in the left anterior descending artery (LAD) and an intermediate branch, and the proximal end of the fistula was closed. The CAF was then opened in proximity to the two CX side branches supplying the lateral LV myocardium, and a footplate, including both side branches, was cut and grafted onto the harvested vein. Hereafter, the left atrial appendage was removed, and by trans-atrial septum access, a size 33 biological mitral valve (Epic, St Jude Medical) was inserted.

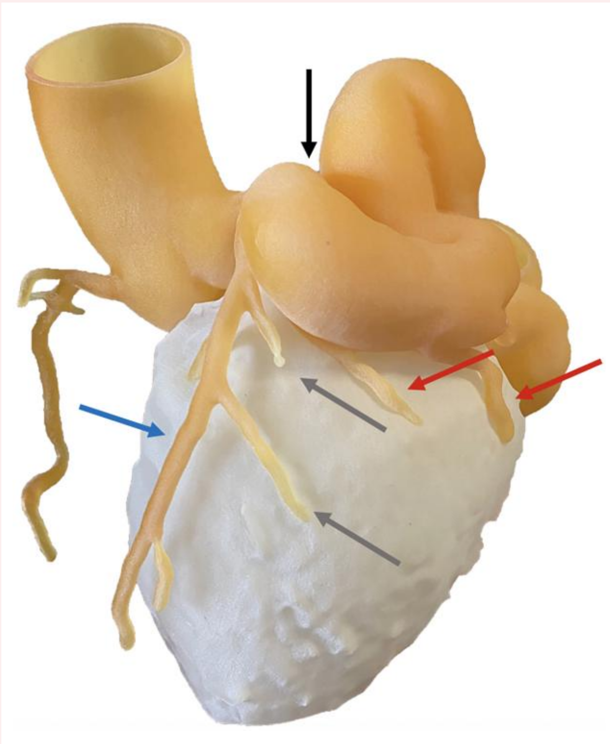


Figure 2 Three-dimensional heart model illustrating the aortic root, the coronary arteries, and the course of the very tortuous and dilated circumflex artery fistula, the dilated coronary sinus, and the left ventricle before surgery. Black arrow pinpoints the hugely dilated circumflex artery. Blue arrow points to the normal-sized left anterior descending artery. Red arrows point to the first and second circumflex artery marginal branches. Grey arrows point to the first and second diagonal branches.

Post-surgery and follow-up

The patient was left with an open sternum due to an intra-operative vasoplegic shock but recovered, and on post-operative Day 3, the sternum was closed. Subsequently, the patient had an uneventful post-operative course and was discharged to a local hospital 17-day post-surgery. The patient has been without symptoms for now 10 months, and CT has shown an open venous graft supplying the obtuse marginal (OM) grafts (*Figure 1E*).

Patient 2

Patient 2 was diagnosed with a CAF after hospital admission for uncharacteristic chest pain and slightly elevated troponins at the age of 47. A coronary angiography revealed a hugely dilated CX artery with drainage through a CAF, connecting to either the CS or the right atrium (RA). Right heart catheterization showed no significant shunting. The patient was discussed at a conference, and it was decided not to do any interventions as the CAF did not cause any sign of coronary stealing or significant shunting and since the patient only had limited symptoms. The patient was advised to take 75 mg of aspirin.

After 13 years of follow-up, the patient developed dyspnoea. Echocardiography showed a significant pericardial effusion, and pericardiocentesis was performed twice within 2 weeks, both times with drainage of large portions (1.7 L) of yellow clear fluid. Despite anti-inflammatory treatment, the patient needed consecutive pericardiocentesis every second week due to recurrence of pericardial effusion. A CT of the thorax and abdomen showed no signs of malignancy, but a massive thrombosis in the terminal part of the CX fistula close to the entrance into the CS, obstructing the drainage to the RA (*Figure 3A* and *B*). It was proposed that the pericardial effusion was caused by the hydrostatic pressure in the massively dilated fistula in combination with the thrombotic sub-occlusion of the CS ostium. Anticoagulation therapy with heparin was started but had no effect within 2.5 months.

At a following conference, the surgeons judged that the CAF did not meet the normal criteria for operation as the patient showed no sign of angina or coronary steal phenomena and there was an uncertainty as to whether an operation with ligation of the fistula would lead to any improvement to his tendency of pericardial effusion. A 3D print of the heart was made to assess the advanced coronary anatomy and possible treatment options.

Three-dimensional print

The 3D-printed heart model (*Figure 3C*) was created as described for Patient 1. Based on the model, the surgeons judged that the CX could be ligated proximally and the two OMs and the ramus descending posterior (RDP) from the CX fistula could be reached by a venous graft. It was found plausible that the high pressure in the dilated CS and coronary veins could be the genesis to the recurrent pericardial effusion by means of transmural filtration. Percutaneous treatment was also discussed, but the highly dilated thin-walled CS was judged to be too fragile. Hence, the patient was admitted for surgery with a vein graft to the three CX branches.

Surgery

The first surgical steps were performed as in Patient 1. Following ECC, the RA and the terminal part of the dilated CX fistula were opened to establish contact between the obliterated CS and the RA. From the RA, the massive thrombus was palpable, and the thrombus material was removed. The terminal part of the fistula was cut out as a footplate and attached end to end to the CS. A vein graft was jumped to the first OM (OM1), which was ligated proximally. The OM 2 was not identifiable. The left main coronary artery was exposed after the division of the main pulmonary artery, and the CX fistula was ligated ostially. Cardiopulmonary bypass was weaned without problems. Post-operative trans-oesophageal echocardiogram (TOE) showed good bi-ventricular function, no shunts, and mild mitral regurgitation. Due to retrograde flow from the first CX marginal branch, the patient was re-operated the day after with further ligation of the upstream part of the concerned CX branch.

Post-surgery and follow-up

After the re-operation, the patient had an uneventful course and was discharged on post-surgery Day 8. Shortly after, the patient was in NYHA Class I. A post-operative CT scan (*Figure 3D*) confirmed an optimal surgery result with an open vein graft with no pericardial effusion, no CS dilation, or thrombus. Repeated echocardiograms after discharge have been without pericardial effusion in a follow-up time of now 1 year.

Discussion

We report two cases of large, tortuous coronary CX fistulas draining into the CS. Both patients were followed from the time of diagnosis until surgery in a time period of more than 10 years. Both had complex CAF anatomy that was difficult to assess on 2D sliced CT images. Therefore, it was uncertain whether surgery was achievable, and a conservative strategy was initially chosen due to a low symptom burden and high surgical risk. Hence, both patients were followed, until they

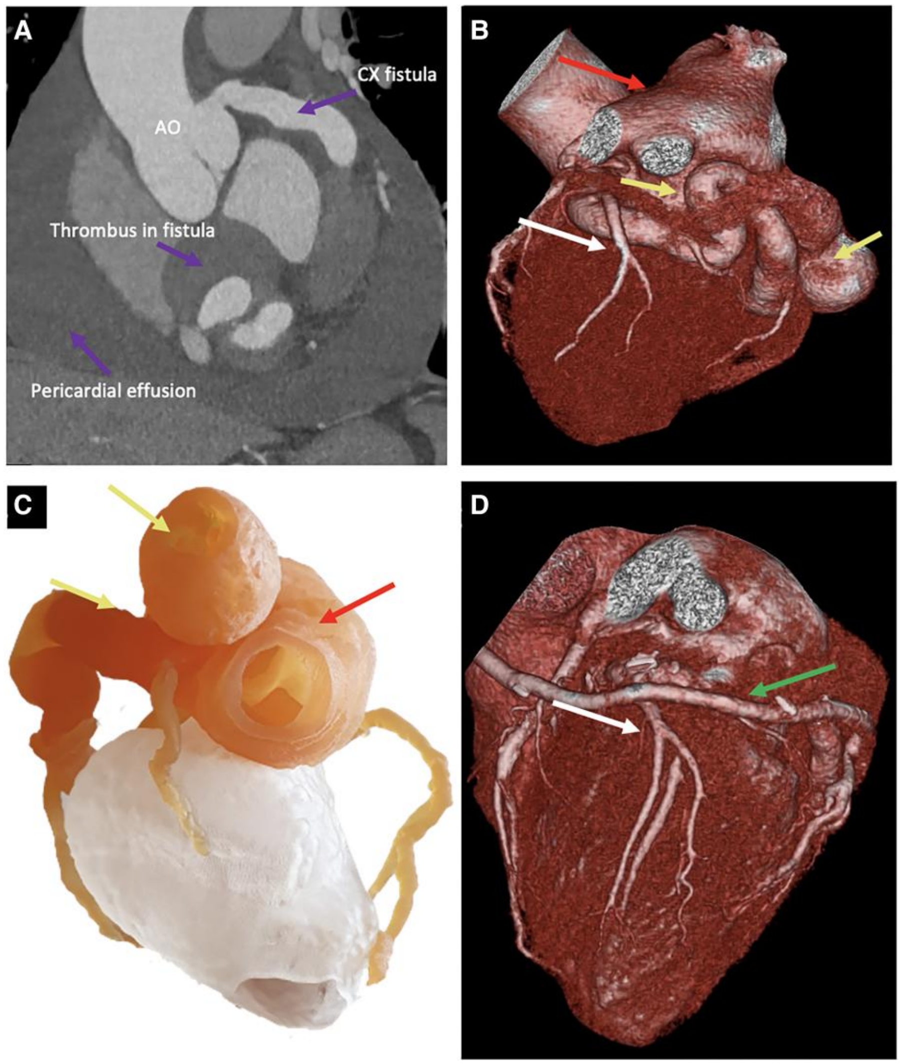


Figure 3 (*A*) Computed tomographic scan before surgery illustrating the fistula and the thrombus in the terminal part of the fistula. (*B*) Computed tomographic scan before surgery illustrating the circumflex artery fistula (yellow arrows) and the obtuse marginal (white arrow). (*C*) Three-dimensional heart model illustration of the coronary arteries, the circumflex artery fistula (yellow arrows), the dilated coronary sinus (red arrow), and the left ventricle before surgery. (*D*) Computed tomographic scan after the surgery showing the venous bypass graft (green arrow) to the obtuse marginals (white arrow).

developed serious progression of symptoms. Patient 1 developed heart failure and severe mitral regurgitation due to increasing fistula dilatation and shunting leading to LV volume overload and coronary steal causing myocardial ischaemia, also affecting the papillary muscle and thereby the mitral valve. Patient 2 had recurrent pericardial effusion, assumably due to compromised venous drainage through the CS.

The reason for the long follow-up, until significant deterioration for these patients, could partly be a consequence of a general reluctance to perform surgery in patients with CAFs, as the very complex anatomy compromises the surgical planning.[6] There is no clear-cut definition when to use 3D prints. Cardiac CT images are visualized on 2D screens, which limits the depth perception and understanding of the true 3D nature of the data. Although surgeons are used to translate clinical CT images into an expected anatomical view of the surgical field, this ability will be challenged by extremely difficult pathoanatomic conditions as in the case of giant coronary fistulas. Under these conditions, an in-hand model will improve the guidance of the surgeon on how to gain access to relevant structures and how to locate the coronary branches from the fistula that needs bypass grafting. Both our patients had very complex pathoanatomic cardiac conditions. Thus, optimal pre-operative investigations were needed for the preparation of a surgical plan that could potentially reduce the risk of the operation. In Case 1, heart transplantation was discussed as an option, but the 3D model assured the surgeon that surgical treatment of the fistula would be feasible.

In Case 2, the thrombosed fistula compromised the drainage of the CS, which was considered to be the cause of the recurrent pericardial effusion. The 3D model convinced the surgeon that the thrombotic segment could be accessed and that the coronary arteries from the fistula could be reached with a bypass graft.

Others have reported a clinical value of the usage of 3D printing in the assessment of CAFs; both Zhang *et al.*[7] and Aroney *et el.*[8] reported in their case reports that the use of 3D printing improved understanding of the pathology for both staff and patients as well as a better preoperative planning.

CAFs and CHD are often associated with complex and unique geometry that can be difficult to appreciate sufficiently from 2D sliced CT images. Hence, 3D-printed heart models may play a key role to provide a more comprehensive understanding of the pathology and offer a unique possibility for personalized treatment.[9]

In a study by Valverde *et al*., 40 patients with complex CHD had a patient-specific 3D-printed model performed, and the best surgical approach was re-defined in 47.5% of the cases.[10] These findings are in line with the experience gained from our two presented cases. Further, a possible benefit of an improved surgical planning is reduced procedural time,[8] which could potentially translate into a reduced perioperative risk. Until now, CT-based 3D printing has primarily been used in isolated cases of CAFs and cases of CHD, and the small sample size in our case report limits the generalizability of our findings. However, a more systematic use of 3D printing in patients with CAF could lead to earlier intervention before clinical progression, as the models may offer a better assessment of the treatment options. Prospective studies using 3D printing more systemically are therefore wanted in order to assess the possible clinical outcome of implementing this technique.[5]

The clinical utility of CT-based 3D printing relies highly on good image quality, since poor images will translate into a less accurate model, which could potentially influence the patient management. However, patient preparation and scanner technology have improved massively the last decades ensuring an overall high image quality in most cardiac CT centres. In a review of 158 non-cardiac studies, the main concerns of 3D printing were related to precision, preparation time, and costs.[10] However, in a recent review of studies using 3D printing in CHD, the accuracy of the models was found to be high, although the accuracy was only reported in 6 out of 28 studies.[4] More larger studies are therefore needed to confirm the accuracy of 3D-printed heart models in different materials. In the same review, the cost to produce the life-sized models ranged from 55 to 810 USD and the segmentation time ranged from 0.5 to 12 h (average was 3.5 h). The cost of 3D printing is difficult to extrapolate from case series since it will depend on model size, materials, and whether in-hospital printing is available. However, the anticipated increased use of 3D printing in cardiology with the introduction of in-hospital 3D-printing laboratories should reduce cost in the future. The segmentation time of 2 h in our cases was relatively low, which is assumably related to the more automatic software solution used.[4,11]

Virtual reality (VR) could potentially also be usable in the preoperative planning of complex anatomy.[12] This advanced method would, however, require extensive training of the cardiac surgeons and for most surgeons an in-hand CT-based 3D model would probably be more relatable to the pathoanatomical findings in the surgical field. However, in experienced hands, 3D VR could be an alternative to 3D printing and could potentially reduce the time related to the preoperative planning by avoiding the relatively long printing time. In this way, 3D VR could potentially be used in more acute/sub-acute settings compared with 3D printing.

Conclusion

In conclusion, we have presented two cases where 3D-printed models influenced the surgical plan in patients with giant CAFs and 3D printing can therefore challenge how we currently image, plan, and perform cardiovascular interventions in complex cases.

Lead author biography

Mads Ørbæk Andersen is a medical doctor and PhD student with high interest in cardiology innovation.

Consent

Both patients have given their consent to this report and have signed the EHJ consent form in accordance with COPE guidelines.

Conflict of interest: None of the authors have any conflics of interest to declare.

Funding: No funding was obtained.

Data availability

Data is available upon reasonable request and permission.

References

1. Yun G, Nam TH, Chun EJ. Coronary artery fistulas: pathophysiology, imaging findings, and management. *Radiographics* 2018;**38**:688–703.
2. Lim JJ, Jung JI, Lee BY, Lee HG. Prevalence and types of coronary artery fistulas detected with coronary CT angiography. *AJR Am J Roentgenol* 2014;**203**:W237–W243.
3. Abdelaty AM, Singh A, McCann GP. Unusual presentation of right coronary artery fistula. *BMJ Case Rep* 2017;**2017**:bcr-2017-220424.
4. Lau I, Sun Z. Three-dimensional printing in congenital heart disease: a systematic review. *J Med Radiat Sci* 2018;**65**:226–236.
5. Wang C, Zhang L, Qin T, Xi Z, Sun L, Wu H, *et al.* 3D printing in adult cardiovascular surgery and interventions: a systematic review. *J Thorac Dis* 2020;**12**:3227–3237.
6. Qureshi SA. Coronary arterial fistulas. *Orphanet J Rare Dis* 2006;**1**:51.
7. Zhang J, Ma W, Zhang W, Kong Y. Three-dimensional printed models-guided surgical repair for recurrent coronary artery fistula. *Ann Thorac Surg* 2019;**107**:e161–e163.
8. Aroney N, Markham R, Putrino A, Crowhurst J, Wall D, Scalia G, *et al.* Three-dimensional printed cardiac fistulae: a case series. *Eur Heart J Case Rep* 2019;**3**: ytz060.
9. Yoo SJ, Hussein N, Peel B, Coles J, van Arsdell GS, Honjo O, *et al.* 3D modeling and printing in congenital heart surgery: entering the stage of maturation. *Front Pediatr* 2021;**9**:621672.
10. Martelli N, Serrano C, van den Brink H, Pineau J, Prognon P, Borget I, *et al.* Advantages and disadvantages of 3-dimensional printing in surgery: a systematic review. *Surgery* 2016;**159**:1485–1500.
11. Bateman MG, Durfee WK, Iles TL, Martin CM, Liao K, Erdman AG, *et al.* Cardiac patient-specific three-dimensional models as surgical planning tools. *Surgery* 2020; **167**:259–263.
12. Sadeghi AH, Bakhuis W, Van Schaagen F, Oei FBS, Bekkers JA, Maat A, *et al.* Immersive 3D virtual reality imaging in planning minimally invasive and complex adult cardiac surgery. *Eur Heart J Digit Health* 2020;**1**:62–70.