JACC: CASE REPORTS VOL. 16, 2023

CASE REPORT

ADVANCED

CLINICAL CASE

3D Printed Model-Guided Neonatal Transcatheter Closure of Left Main Coronary Artery-to-Right Ventricle Fistula

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ABSTRACT

We report on a 2-week-old infant with huge left main coronary artery-to-right ventricular outflow tract fistula causing myocardial ischemia due to global coronary steal who was successfully submitted to percutaneous closure guided by a 3-dimensional-printed model using a duct-occluder vascular plug. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2023;16:101869) © 2023 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

Coronary artery fistula (CAF) is an uncommon congenital anomaly affecting up to 0.5% of the population. It is defined as an abnormal communication between the coronary artery bed and a cardiac chamber or intrathoracic great vessel without interposition of the capillary network.[1,2] Although most patients are asymptomatic, this malformation may cause clinical symptoms and cardiovascular complications at any age, including myocardial ischemia, heart failure, infective endocarditis, and rupture with resulting sudden death.[3] Treatment of this malformation may be surgical or transcatheter closure based on echocardiographic, angiographic, or computed tomography (CT) scan evaluation. However, these diagnostic methods often fail to detail the anatomy of the coronary malformation and its relationship with adjacent structures, mainly in small-size pediatric patients. In these challenging anatomic settings, 3-dimensional (3D) printing of cardiac models could be a useful tool in

LEARNING OBJECTIVES

- To highlight that early recognition and closure of large symptomatic CAFs are crucial to achieving normal cardiac functions and adequate growth.
- To show the usefulness of the 3D printing technology in transcatheter treatment of challenging coronary artery anomalies in terms of preprocedure planning and device selection.
- To show the impact of this approach in improving outcome of "complex" transcatheter intervention in small infants.

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

Manuscript received January 22, 2023; revised manuscript received March 27, 2023, accepted March 31, 2023.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2023.101869

ABBREVIATIONS AND ACRONYMS

CAF = coronary artery fistula

CT = computed tomography

3D = 3-dimensional

planning the therapeutic approach. It allows to finely detail course, relationship with adjacent structures, and site of drainage of the CAF, so potentially improving the procedure and patient outcome.[4,5,6]

Here, we report on a case in which 3D printing technology was crucial in transcatheter closure of a "complex," symptomatic CAF in neonatal age.

HISTORY OF PRESENTATION

A 2.9-kg, 2-week-old infant was referred to our department because of cardiac murmur and failure to thrive.

PAST MEDICAL HISTORY

No significant problem had been reported during the prenatal and perinatal periods, and the symptoms ensued during the first few days of life.

DIFFERENTIAL DIAGNOSIS

At clinical examination, a harsh continuous murmur at midsternal level was found. Faint intercostal and subcostal retractions were evident. Patent arterial duct, aortopulmonary window, and coronary-cameral fistula were the potential differential diagnoses.

INVESTIGATIONS

Routine laboratory tests showed high level of N-terminal pro-B-type natriuretic peptide (51,482 ng/dL) and high-sensitivity troponin T (280 ng/dL). Chest x-ray film showed mild cardiac enlargement and mild increase of pulmonary vascular markings. Electrocardiography showed nonspecific ST-T changes in anterior-inferior leads. At echocardiography, mild left cardiac chamber overload with normal ventricular function was imaged. At color Doppler analysis, a significant, continuous flow into the right ventricular outflow tract was imaged, with significant dilatation of both right and left coronary arteries and a prominent Vieussens' ring (**Figure 1**). Thus, the diagnosis of CAF with "complex" anatomy was suspected. At CT scan evaluation, the coronary artery anatomy was better detailed, showing a huge coronary fistula (5.5 × 3.7 mm) from the left main coronary artery to the right ventricular infundibulum that caused significant steal also from the right coronary artery via the Vieussens' ring (**Figure 2A**).

MANAGEMENT PLAN

At the heart team discussion, the fistula was considered suitable for early percutaneous closure. This approach was deemed more favorable than medical management of heart failure and delayed transcatheter closure. However, to effectively plan the intervention in terms of vascular access (ie, femoral vs jugular vein) and procedure steps, a CT scan-based 3D model of the fistula was printed (**Figures 2B and 2C**, Video 1). It was used to detail the relationship between the Vieussens' ring and the drainage site of the fistula, mimic ex vivo all steps of the procedure, and finally, test different occluding devices. At cardiac catheterization, the left-to-right shunt across the fistula was mild, so precluding clear visualization of the site and size of the fistulous drainage site by either aortic or right ventricular angiography (**Figures 3A to 3C**). Selective coronary angiography was not performed due to the small size of the patient, and aortic angiography clearly imaged just the Vieussens' ring. However, the fistula was successfully probed from the right ventricle on the basis of the anatomic details obtained from the printed 3D model and angiographically imaged using an arteriovenous circuit (**Figure 3D**). As anticipated, a duct-occluder vascular plug was chosen to occlude the fistula and deployed by a dedicated 4-F catheter from the venous site, without any interference with the left coronary artery. Complete occlusion of the fistula was confirmed at control aortic angiography (**Figures 4A and 4B**, Video 2). The procedure was uneventful, and the infant was discharged in good clinical conditions in a few days.

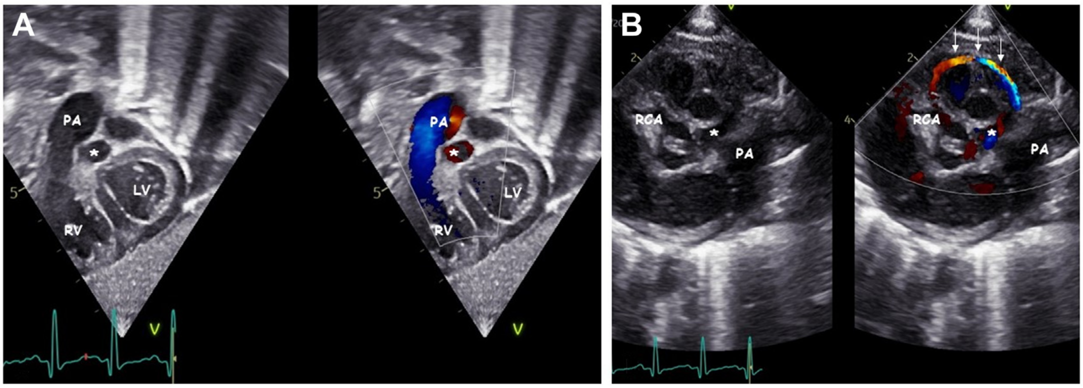
FOLLOW-UP

After the procedure, cardiac failure signs completely reverted in a few days without pharmacologic therapy. Echocardiographic and CT scan evaluations confirmed the complete and successful closure of the fistula (**Figures 4C and 4D**, Video 3).

DISCUSSION

Coronary artery fistulas are often asymptomatic, although they may cause left ventricular enlargement in the case of moderate left-to-right shunt. Symptoms of congestive heart failure and myocardial ischemia are very rare, mainly due to a very large shunt with coronary artery steal.[7] In the neonatal period, this physiopathologic condition is rarely found, mainly in the case of CAF involving multiple coronary branches or left main coronary artery.[7,8] Indeed, the combination of cardiac volume overload due to left-to-right shunt and impairment of coronary blood flow caused by diffuse coronary steal may result in cardiac failure caused by global myocardial ischemia.

FIGURE 1 Echocardiographic Findings of the Coronary Artery Fistula

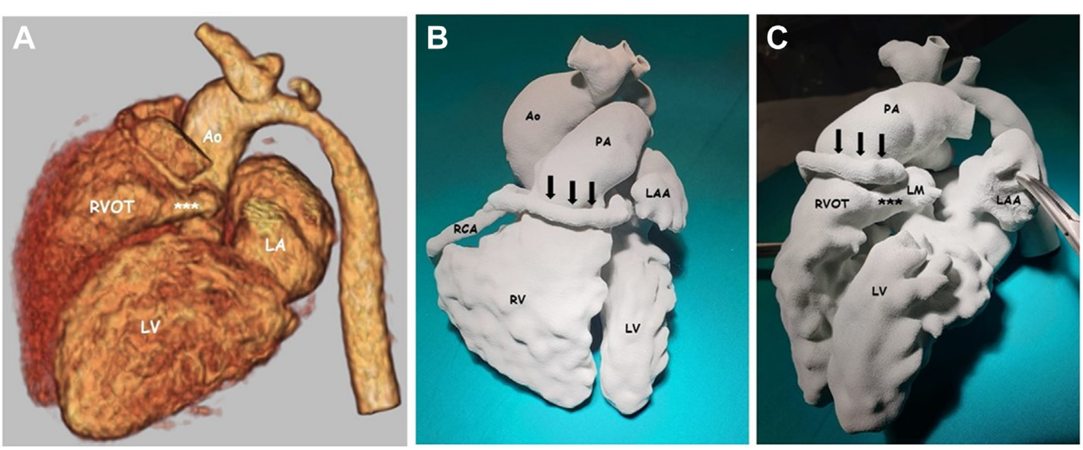


(A) Echocardiographic image in subcostal left oblique view of the site of drainage **(asterisk)** of the left main coronary artery (LMCA) fistula into the right ventricular (RV) outflow tract. Echocardiographic image in parasternal short-axis view showing the dilated right coronary artery (RCA) and Vieussens' ring **(arrows)** draining into the coronary artery fistula **(asterisk)**. LV = left ventricle; PA = pulmonary artery.

Treatment of significant CAFs may be surgical or by transcatheter approach. This latter approach is often challenging in the neonatal age due to the patient size and the anatomic complexity of the malformation. In this setting, 3D technology has recently emerged as an effective tool for preprocedural planning of complex transcatheter interventions.[5,6,9-12] This approach allows to finely detail complex anatomies; simulate ex vivo the planned procedure, selecting the best vascular access and the most suitable occluding device; and prevent potential technical problems and complications.

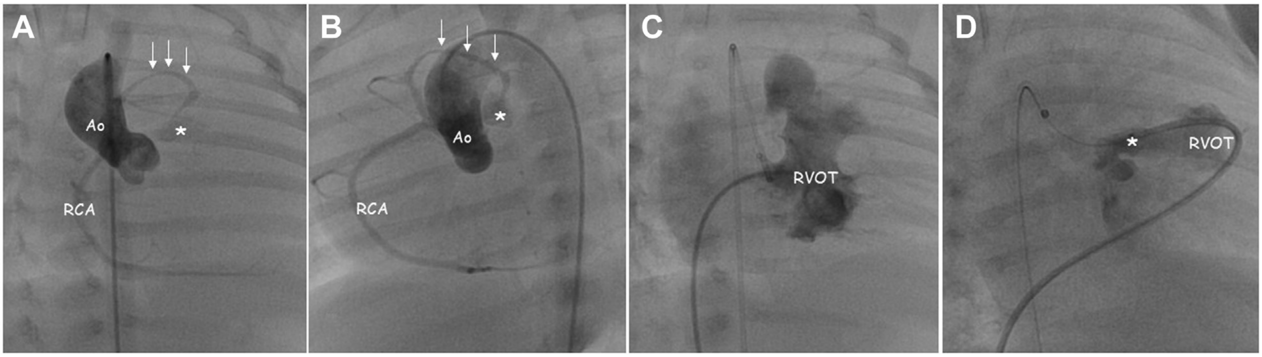
To date, only a few reports of transcatheter treatment of CAF guided by 3D-printed models for have

FIGURE 2 Advanced Imaging of the Coronary Artery Fistula



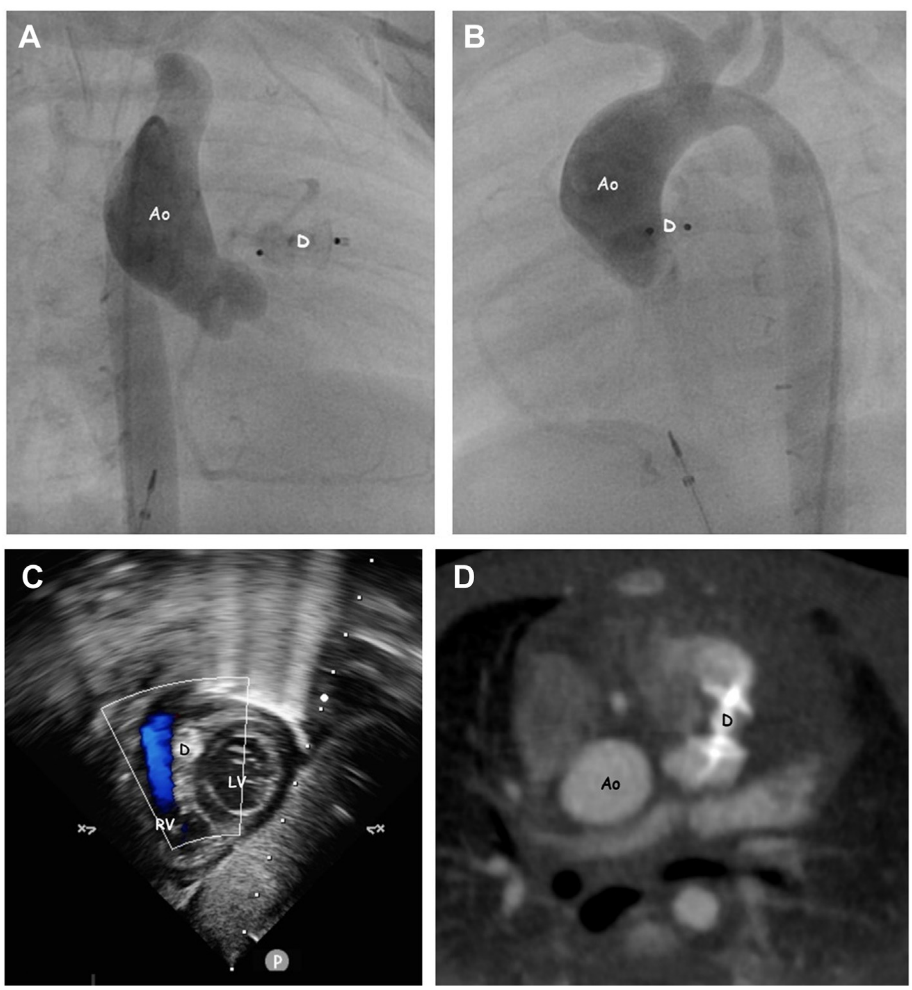
(A) Computed tomography scan 3-dimensional reconstruction of the site of drainage of the coronary artery fistula into the right ventricular outflow tract (RVOT). Printed 3-dimensional model in **(B)** frontal and **(C)** lateral views, showing the dilated Vieussens' ring **(black arrows)** as the steal pathway from the RCA to the drainage site **(asterisks)** of the fistula into the RVOT. Ao = aorta; LAA = left atrial appendage; LM = left main coronary artery; other abbreviations as in Figure 1.

FIGURE 3 Baseline Angiographic Findings of the Coronary Artery Fistula



Both **(A, B)** aortic and **(C)** RV angiographies failed to detail the coronary fistula anatomy. **(D)** However, based on the 3-dimensional-printed model, it was possible to cross and image the site and size of the coronary artery fistula drainage site **(asterisk)**. **Arrows** indicate Vieussens' ring. Abbreviations as in Figures 1 and 2.

FIGURE 4 Final Result After Vascular Plug Deployment Into the Coronary Artery Fistula



Aortic angiography in **(A)** right and **(B)** left oblique views after closure of the fistula with a duct-occluder vascular plug. No residual shunt is imaged at **(C)** echocardiographic and **(D)** computed tomography scan evaluation. Ao = aorta; D = device.

been published in literature.[4,6] They reported adult patients in whom the use of this technology increased feasibility and outcome of the transcatheter approach. These advantages should be even more tangible in the case of small-size neonates in whom echocardiography and CT scan often lack sufficient sensibility to obtain adequate anatomic details. Detailing in advance the anatomy of this malformation may significantly decrease the impact of the interventional catheterization in terms of procedure time, x-ray dose, and contrast medium amount. In our patient, this approach made possible to detail in advance the site of drainage as well as the size and length of the fistulous course in order to test ex vivo the best-occluding device. In addition, it was crucial in probing the fistula because the angiographies either from the aorta or from the right ventricle had failed to image the site of drainage and the morphology of the malformation.

CONCLUSIONS

Technical advances in 3D model technology and devices could be highly beneficial in transcatheter treatment of challenging anatomies, mainly in small infants.

FUNDING SUPPORT AND AUTHOR DISCLOSURES

This work was supported by the 3D VIRTUAL BABY HEART project (2018-2020) funded by the Italian Ministry of Health (grant number: GR-2016-02365072). Dr Santoro has served as a proctor of Abbott, W.L. Gore, and Occlutech, Italy. All other authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS congenital heart defect, coronary vessel anomaly, myocardial ischemia, occluder, percutaneous coronary intervention, 3-dimensional printing

APPENDIX For supplemental videos, please see the online version of this paper.