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CASE REPORT

ADVANCED

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Transcatheter Closure of Left Ventricle to Coronary Sinus Fistula Post-MVR and Septal Myectomy

Rory S. Bricker, MD,[a] Robert A. Quaife, MD,[a] Shiuh-Yung J. Chen, PhD,[a] John C. Messenger, MD,[a] Jacob Hammers, PhD,[b] John D. Carroll, MD[a]

ABSTRACT

This paper describes the case of a patient who developed refractory heart failure due to a fistula from the left ventricle to the coronary sinus that was unintentionally created after a surgical myectomy and mitral valve replacement. Advanced image guidance with a pre-procedure 3-dimensional physical model and intraprocedure echocardiography fusion facilitated transcatheter plugging of the shunt with symptom resolution. (**Level of Difficulty: Advanced.**)

HISTORY OF PRESENTATION

A 71-year-old female presented with progressive ambulatory New York Heart Association (NYHA) functional classes III to IV symptoms of dyspnea, edema, abdominal distention, and severe fatigue refractory to high-dose diuretics.

LEARNING OBJECTIVES

- To identify the rare complication of fistula formation following mitral valve surgery.
- To understand that transcatheter techniques for fistula closure can be safe and effective therapies for a patient at prohibitive surgical risk.
- To integrate advanced imaging into pre-procedure planning and intraprocedural guidance to improve procedural success.

MEDICAL HISTORY

The patient had a history of symptomatic hypertrophic obstructive cardiomyopathy and severe mitral regurgitation with mitral annular calcification (MAC). A surgical septal myectomy and mitral valve replacement operation was technically complex. The anterior mitral leaflet was excised, and a 29-mm Magna Ease (Edwards Lifesciences, Irvine, California) bioprosthetic valve was placed in an atrialized mitral position, lying 2 cm above the mitral annulus, as the annulus was small due to MAC and the risk of atrioventricular disruption was high. Postoperatively, refractory heart failure was incapacitating, despite medical management.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis included heart failure due to paravalvular leak, patient prosthesis mismatch,

From the [a]Department of Medicine, Division of Cardiology, University of Colorado School of Medicine, Aurora, Colorado, USA; and [b]Philips Healthcare, Andover, Massachusetts, USA.
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diastolic dysfunction, pericardial constriction, and intracardiac shunt.

INVESTIGATIONS

On examination, a loud holosystolic murmur was present. Transthoracic echocardiography (TTE) demonstrated a high-velocity Doppler signal in the region of the posterior mitral valve annulus and high-velocity systolic flow from the coronary sinus (CS) into the right atrium (**Figure 1**, Videos 1, 2, and 3).

MANAGEMENT

The heart team agreed that she was prohibitively high risk for repeated surgical intervention. The patient was referred for a transcatheter-based repair.

Procedural planning was completed using cardiac computed tomography angiography (CTA) (**Figure 2**) and subsequent image segmentation with creation of a standard tessellation language (STL) file for rapid prototyping a 3-dimensional (3D) model (Supplemental Figures 1 and 2, **Figure 3**) (1). The procedure was performed with the patient under general anesthesia with fluoroscopic and TEE guidance. A left ventriculogram confirmed severe left-to-right shunting with complete opacification of the CS and right atrium (**Figure 4C,** Video 4). Real-time echocardiographic-fluoroscopic fusion imaging (EchoNav, Philips, The Netherlands) (2) was used to overlay a marker denoting the fistula origin onto live fluoroscopic images to facilitate fluoroscopy-based guidance to cross the defect from the right atrium (Videos 5, 6, and 7).

The fistula was successfully crossed through the right internal jugular venous approach using an 8-F Agilis deflectable guide catheter (Abbott, Abbott Park, Illinois) to engage the CS, with a telescoped 0.014-deflectable tip Venture catheter (Teleflex Inc., Minneapolis, Minnesota) advanced into the CS to direct the balance middle weight wire (Abbott) across the defect. The wire was advanced into the aortic arch.

A 90-cm 0.035 Navicross catheter (Terumo Interventional Systems, Terumo, Tokyo, Japan) was advanced over the balance middle weight wire to the aortic root, the wire was exchanged for an Amplatz Extra Stiff (ES) (Cook Medical, Bloomington, Indiana), and a 6-F × 90-cm shuttle sheath (Cook Medical), was advanced into the aortic root. The dilator was removed, and a 0.014-Grand Slam (Abbott) was advanced to the descending aorta as a "safety wire" to maintain position across the defect adjacent to the working wire. The shuttle sheath was then removed and re-advanced across the fistula into the left ventricle over only the Amplatz ES wire (**Figure 4A**).

A 10-mm Amplatzer II vascular plug (AVP II) (Abbott Vascular) was deployed through the shuttle sheath with the distal lobe retracted against the fistula orifice in the ventricularized portion of the left atrium with the remaining segments deployed in the CS (Video 8). Venography demonstrated complete occlusion of the CS (**Figure 4B,** Video 9). The device was recaptured with the shuttle sheath and exchanged for an 8-mm AVP 4 which was unstable and pulled through the defect.

The "safety wire" was then used to reposition the delivery sheath across the defect. Next, an 8-mm AVP II was deployed with the distal disc flush against the ventricularized left atrium and the remaining two lobes deployed in the CS (Video 10). Left ventriculography demonstrated minimal residual left-to-right shunting (**Figure 4D,** Video 11), and TEE images confirmed a patent CS with decreased systolic flow. The AVP device was released and stable within the tract.

ABBREVIATIONS AND ACRONYMS

AVP = Amplatzer vascular plug

CS = coronary sinus

CTA = computed tomography angiography

LV = left ventricle

MAC = mitral annular calcification

NYHA = New York Heart Association

TEE = transesophageal echocardiogram

TTE = transthoracic echocardiogram

VSD = ventricular septal defect

DISCUSSION

Intracardiac fistula formation following mitral valve replacement is a rare surgical complication. Left ventricle-to-coronary sinus (LV-CS) fistula is a more rarely reported subtype (3-7). Multiple mitral valve surgeries, excessive debridement of the mitral valve annulus, or injury to the posterior wall are believed to increase the risk of fistula development. Most reported cases have required surgical repair (3), whereas 1 limited case report described closure of an LV pseudoaneurysm to CS fistula by percutaneous placement of 2 ventricular septal occluders (8).

The authors present a unique case of fistula formation from "ventricularized" atrial tissue into the CS following mitral valve replacement that was successfully treated with an Amplatzer vascular plug. The driving pressure producing that massive shunt was the high ventricular pressure despite the origin of the fistula being in the left atrium. To the authors' knowledge, that is the first case report of LV-CS fistula treated percutaneously. An alternative approach of placing a covered stent in the CS across the fistula entry was considered, although not attempted due to the concern of stent thrombosis.

This case demonstrates the feasibility, safety, and efficacy of a percutaneous LV-CS fistula closure.

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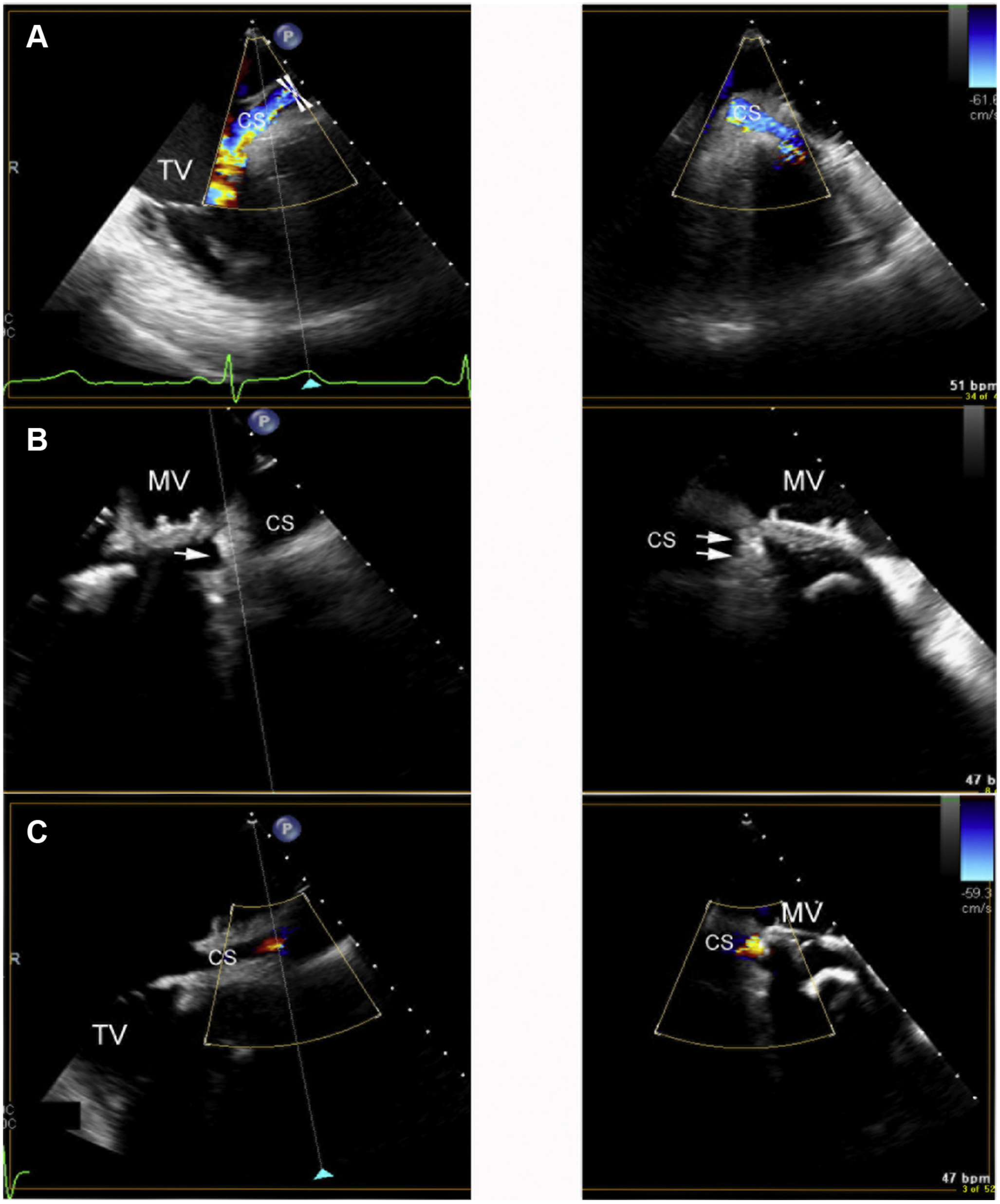
FIGURE 1 **Transesophageal Echocardiography**



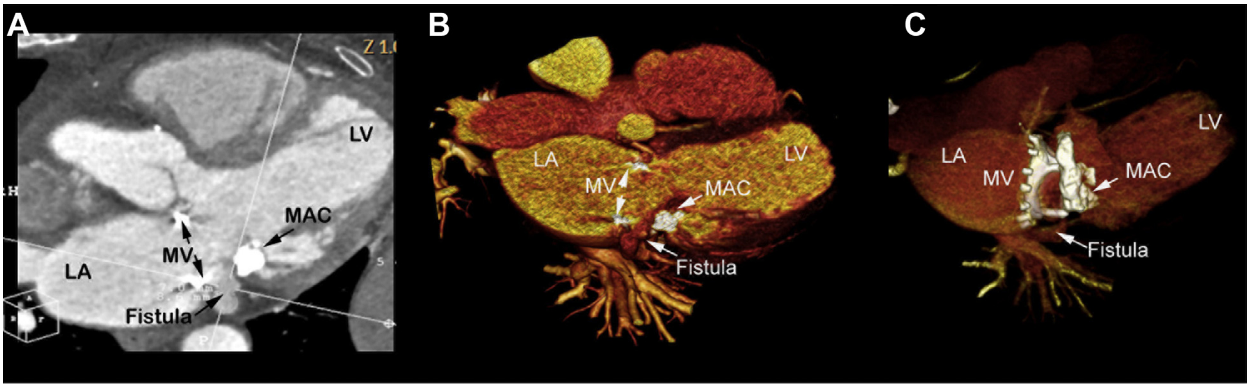
(A) Pre-closure low mid esophageal right ventricle biplane view demonstrates a high-velocity jet through the CS into the right atrium. **(B)** Amplatzer occluder device in the CS **(arrows)**. **(C)** Post-closure low mid esophageal right ventricle biplane view demonstrating a patent CS with low residual fistula jet. CS = coronary sinus.

Compared to reoperation, the endovascular approach decreased recovery time and cost, while avoiding the high surgical risk associated with recent sternotomy. The long-term risks and durability of percutaneous LV-CS fistula closure are unknown, although procedures following a similar technique for ventricular septal defect (VSD) occlusion have demonstrated favorable efficacy, procedural safety, and short-term clinical outcomes (9,10).

FOLLOW-UP

After a successful endovascular closure, the patient's heart failure symptoms resolved with markedly

FIGURE 2 Computed Tomography



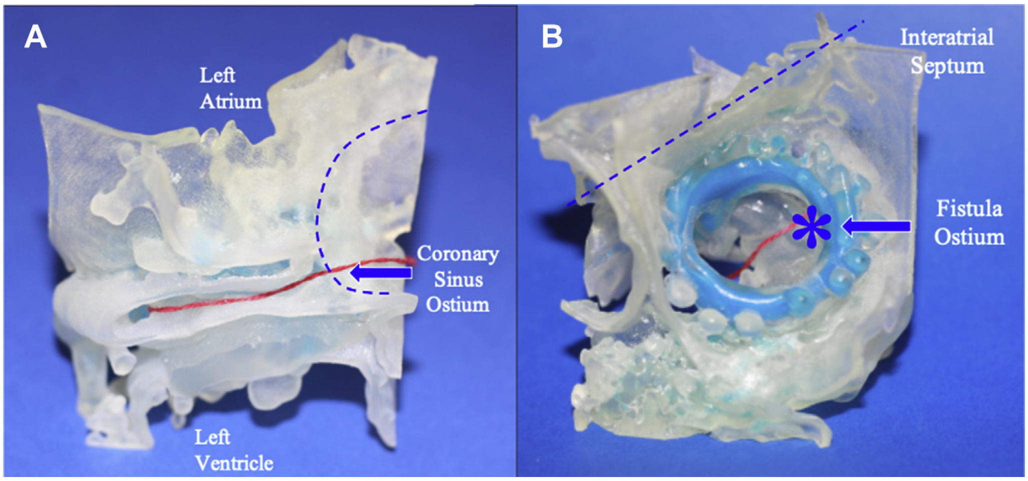
A multiplanar planar reformat (MPR) technique was used to facilitate defect evaluation. The defect size was estimated at 8.35 x 10 mm in diameter, 61 mm^2 in area, and 28.6 mm in perimeter. **(A)** A cross section of the CS parallel to the fistula with flow demonstrated between the LV and CS. **(B)** A volume rendering of the segmented CT data set parallel to the long axis of CS and **(C)** perpendicular to the CS at the connection illustrates the spatial relationship among the fistula. CS = coronary sinus; CT = computed tomography; LA = left atrium, LV = left ventricle; MAC = mitral annular calcification.

improved self-reported quality of life, decreased diuretic requirements, and no hospital admissions 20 months following the procedure. Imaging by cardiac CT and TTE demonstrated device stability with no left-to-right shunt through the original defect (Videos 12 and 13).

CONCLUSIONS

Fistula formation with intracardiac shunting is a rare complication following mitral valve replacement that has conventionally required surgical treatment. Transcatheter fistula closure using a vascular plug appears to be a safe

FIGURE 3 3D Printed Model



3-dimensional (3D) model view of the fistula with CS external wall cut away **(A)** and from the right atrial vantage point or surgeon's view **(B)**. The **dotted line** represents the interatrial septum. The **red string** denotes the course of the fistula from the LV ostium **(*)** to the right atrium. Abbreviations as in Figure 2.

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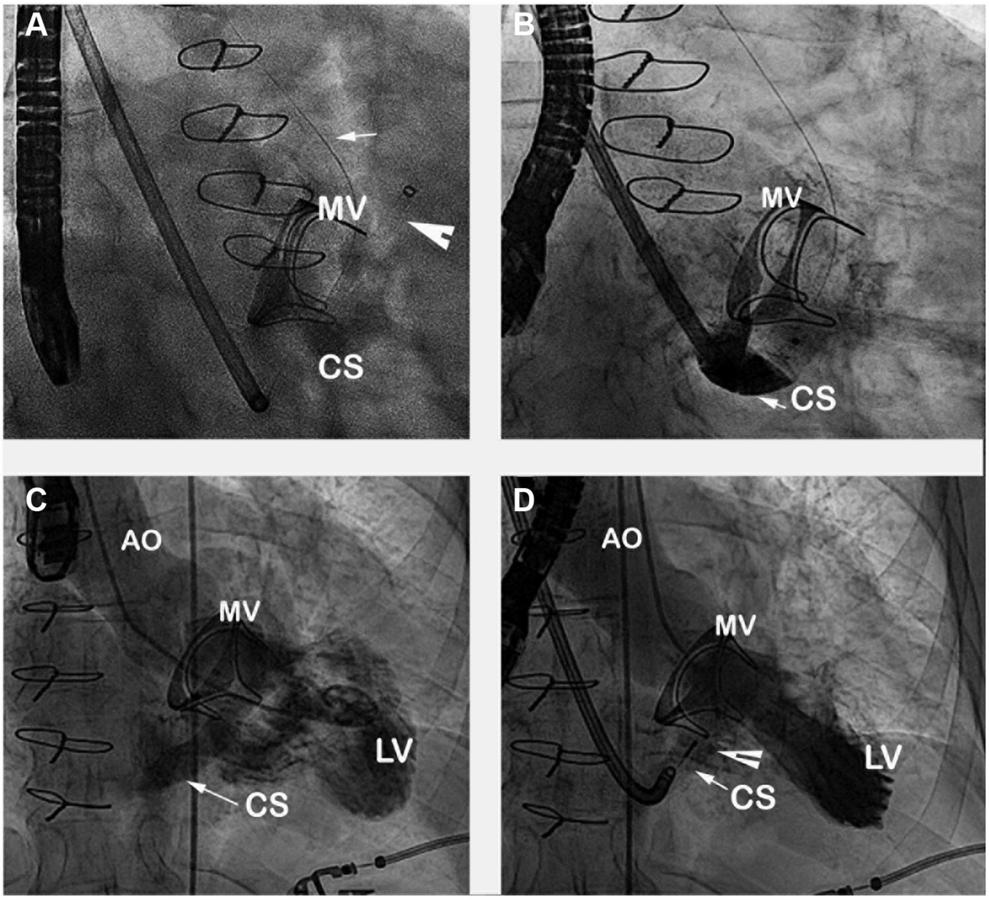
FIGURE 4 Fluoroscopy and Angiography



(A) A shuttle sheath used for device delivery **(large arrow)** is passed through an Agilis sheath (Abbott) through the CS and across the defect. A second wire **(small arrow)** was placed to maintain position. **(B)** Angiography with contrast injection into the CS demonstrates complete occlusion by the 10-mm AVP II. **(C)** Pre-closure left ventriculography demonstrates severe left-to-right shunt through the CS. **(D)** Final left ventriculography with 8-mm AVP II device in place, demonstrating minimal residual shunt through the device. AVP = Amplatzer vascular plug.

and effective treatment strategy for a patient at high risk for repeated surgery. Preprocedural planning with creation of a 3D model and intraprocedural fusion imaging are key components to this approach.

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ADDRESS FOR CORRESPONDENCE: Dr. Rory Bricker, University of Colorado School of Medicine, Mail Stop B 132, Leprino, 12401 East 17th Avenue, Aurora, Colorado 80045, USA. E-mail: rory.bricker@cuanschutz.edu.

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KEY WORDS fistula, fusion, imaging, percutaneous, plug, structural

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