

MINI-FOCUS ISSUE: COMPLICATIONS

INTERMEDIATE

CASE REPORT: CLINICAL CASE

Percutaneous Closure of an Iatrogenic Inlet Ventricular Septal Defect in an Atrioventricular Canal



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ABSTRACT

A 45-year-old woman with a partial atrioventricular canal presented with an iatrogenic interventricular shunt after implantation of a mechanical mitral prosthesis. The shunt was occluded percutaneously with an Amplatzer Duct Occluder II. This is the first reported percutaneous closure of a ventricular septal defect in an atrioventricular canal defect. (Level of Difficulty: Intermediate.) (J Am Coll Cardiol Case Rep 2020;2:911-4) © 2020 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/>).

HISTORY OF PRESENTATION

A 45-year-old woman with a partial atrioventricular septal defect (AVSD) with an ostium primum atrial septal defect (ASD) and mitral cleft presented with subacute dyspnea at the outpatient clinic 2 months after she had a mitral St. Jude prosthesis (St. Jude Medical Master Series, St. Paul, Minnesota) implanted. Clinical examination revealed a rough parasternal murmur.

MEDICAL HISTORY

The patient had first surgery at age 27 years, with closure of the ASD and suture of the mitral cleft,

followed 5 years later by a second surgery for a new mitral plasty and tricuspid annuloplasty.

Six years later, she underwent pacemaker implantation for complete atrioventricular block. Recently, she developed recurrence of mitral regurgitation, indicating the need for mitral replacement with a St. Jude Medical Master Series mechanical prosthesis. The post-operative period was difficult and was complicated by pneumonia and poorly tolerated atrial fibrillation.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis for this murmur was a paravalvular leak, a ventricular septal defect (VSD), or tricuspid regurgitation.

LEARNING OBJECTIVES

- To understand the anatomy and mechanism of the shunt in an unusual VSD. This implies a good knowledge of the congenital disease anatomy.
- To consider the off-label use of more appropriate devices for closure of a shunt in special anatomies.

INVESTIGATIONS

Echocardiography showed the presence of an interventricular shunt located beside the annulus of the mechanical mitral valve. Heart catheterization confirmed the relevance of the shunt, with a Qp/Qs calculated at 1.7.

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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 *JACC: Case Reports* [author instructions page](#).

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**ABBREVIATIONS
AND ACRONYMS****ASD** = atrial septal defect**AVSD** = atrioventricular septal defect**TEE** = transesophageal echocardiography**VSD** = ventricular septal defect**MANAGEMENT**

After heart team discussion because of multiple operations and complications after the last surgery, we decided to perform a percutaneous closure, despite the lack of data on this type of VSD.

The procedure was performed under general anesthesia to allow for continuous guidance with transesophageal echocardiography (TEE). The aortic valve was easily passed with a 0.035-inch guidewire and a 5-F AR II catheter (Cordis, Santa Clara, California) through the left femoral artery. This one has a 90° angulation well adapted, in our experience, to the crossing of a VSD. The VSD was crossed by a retrograde approach, and the AR II catheter was then positioned in the pulmonary artery. A 5-F multipurpose catheter was placed in the pulmonary artery through the right femoral vein. A 30-mm multi-snare was used to capture the first retrograde guidewire in the pulmonary artery and to pull it to the right femoral vein, which achieved an arterial-venous loop ([Video 1](#)).

Using the anterograde approach, the 6-F delivery sheath could not pass the VSD because of a difficult path through the tricuspid subvalvular apparatus. We thus changed to a retrograde approach, repositioning the AR II catheter in the venous part of the looping guidewire. A JR4 5-F guiding catheter (Medtronic, Minneapolis, Minnesota), chosen for its curvature that allowed easy passage through the tunnel of the VSD, was introduced through the left femoral artery and was used for angiography of the VSD, which allowed for an exact measurement ([Figure 1](#), [Video 2](#)). The VSD was 4 mm in width and 3 mm in length. The delivery sheath, introduced through the left femoral artery, was driven on the looping guide into the right ventricle until contact was made with the AR II catheter.

We chose a St. Jude Amplatzer Duct Occluder II (ADO II) 6/4 mm, because it seemed to be the most adaptable to the anatomy of the defect, mainly due to the adaptability of the waist. Delivery of the distal and proximal disks was easy ([Video 3](#)), without any difficulty in unfolding the right disk near the tricuspid subvalvular apparatus and no interference with the mitral prosthesis. We confirmed good attachment by mobilization.

Ventriculography and TEE confirmed the occlusion of the shunt and absence of interference of the device with the mitral prosthesis or tricuspid valve ([Video 4](#)). The device was then released ([Figure 2](#)). The patient was monitored for 24 h and then discharged.

DISCUSSION

Despite good acute surgical results with low mortality and complication rates, re-operations in partial AVSD are frequent, essentially because of mitral insufficiency recurrences or left outflow tract obstructions (1). Surgery for residual shunts has a Class I recommendation when the shunt is ≥ 1.5 without pulmonary hypertension (2).

The anatomy of AVSD is complex. In the partial form, there is a unique atrioventricular annulus with an anterior bridging leaflet attached on the scooped-out ventricular septum (3). Thus, even with no shunt at the ventricular level, the inlet septum will not be normal, and a defect will be present but hidden by the attachment of the anterior bridging leaflet on the septal crest. In the present case, the mechanism of development of the shunt after mitral replacement probably involved a change in geometry, perhaps followed by a leave-in suture point because the prosthesis was sutured onto valvular tissue.

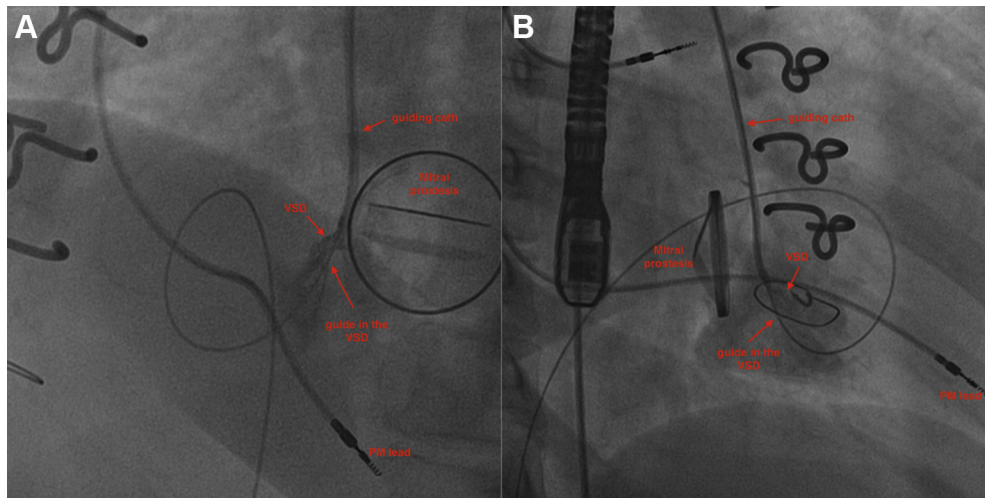
In such a complex anatomy, percutaneous closure has never been attempted, and surgery is current practice. In this case, the high operative risk combined with the technical difficulty and risk for recurrence prompted us to opt for percutaneous intervention. This approach could be attempted with comparatively little risk; surgery could be pursued only in case of failure.

Transcatheter closure of iatrogenic VSDs has been described, typically after aortic valve replacement surgery (4), and more recently, following transcatheter aortic valve implantation. In some of these cases, transcatheter procedures using Amplatzer muscular septal occluders have been successful (5). Iatrogenic VSD following mitral valve replacement in an acquired mitral disease has been reported, with successful occlusion using an ADO II and a retrograde approach (6).

The case we describe here offers many procedural lessons. The anterograde approach, usually advocated in percutaneous VSD closure, could not be applied because of an obstacle in the tricuspid subapparatus. Conversion to a retrograde approach offered easy positioning of the closure device.

All VSD closure devices are dedicated to specific anatomies but are not well suited to unusual anatomies. In many situations, off-label use of non-dedicated devices for VSD closure is necessary as Udink ten Cate et al. (7) recently showed with the use of single-disk duct occluders in membranous VSDs. The off-label use of ADO II for percutaneous closure of membranous VSDs is also common (8).

FIGURE 1 Angiography of the VSD



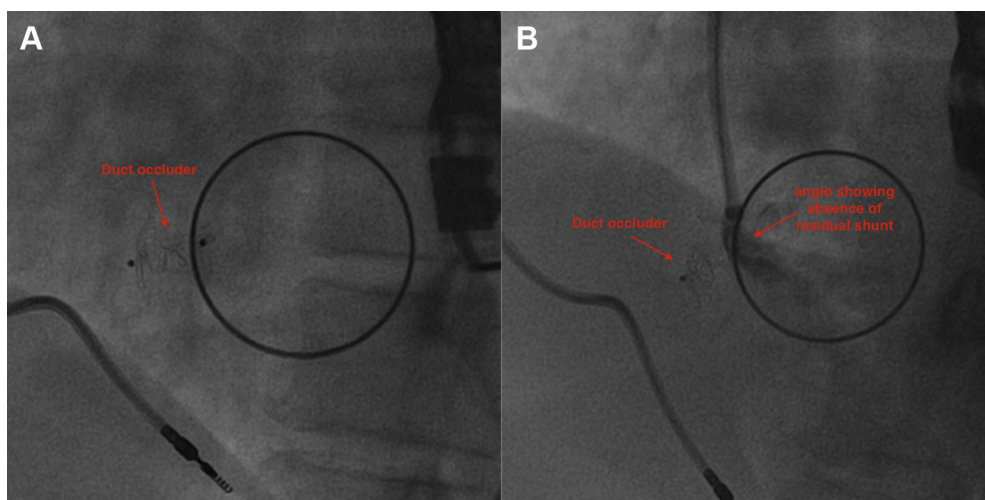
Angiography of the ventricular septal defect (VSD) on the guidewire arterio-venous loop, visualized at 9 h near the mitral St. Jude prosthesis. Note the important loop of the guidewire around the tricuspid valve, which explains the failure of the VSD crossing by the anterograde approach. **(A)** Left anterior oblique 30° incidence and **(B)** right anterior oblique 30° incidence. PM = pacemaker.

The ADO II was of special efficacy in this case. It is a nitinol auto-expandable device dedicated to patent duct percutaneous closure. Its symmetric morphology is suitable for both anterograde and retrograde approaches. We recommend choosing a slightly oversized waist to gain good compression and complete closure of the defect. With the ADO II, the

flexibility of the disks could theoretically reduce the risk of an atrioventricular conduction lesion.

In our case, only angiography could allow for precise measurement of the defect and the appropriate device choice. Peri-procedure TEE is essential to identify interference with the nearest valvular structures and to ensure safe release of the device.

FIGURE 2 Amplatzer Duct Occluder II



(A) Amplatzer Duct Occluder II in its final position after delivery in left anterior oblique at 30°. **(B)** Selective injection beside the left disk confirmed the absence of residual shunting.

FOLLOW-UP

After this procedure, the patient showed spectacular improvement, with no events at 2 years of follow-up. There was no hemolysis documented, which was expected because of the absence of a residual shunt.

CONCLUSIONS

To our knowledge, this case involved the first description of percutaneous closure of an iatrogenic inlet ventricular shunt after mitral prosthesis

implantation in an atrioventricular canal defect. Our case shows that when surgery is risky and technically difficult, percutaneous closure can be attempted even in complex anatomies.

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KEY WORDS congenital heart disease, iatrogenic, percutaneous closure

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