Aorta-to-right ventricle neoshunt closure using an Amplatzer Duct Occluder II device

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Abstract
We report the case of a 22-year-old female patient with complex congenital heart disease and multiple cardiac surgeries who came to our attention for right heart failure and hemolysis 3 years after aortic valve replacement surgery. She was diagnosed with aorta-to-right ventricle fistula and was efficiently treated with retrograde implantation of an Amplatzer Duct Occluder II device using three-dimensional multimodality fusion imaging.

KEYWORDS
Amplatzer Duct Occluder II, aorta-to-right ventricle fistula, device closure

1 | INTRODUCTION

Aorta-to-right ventricle (RV) shunt is a very rare phenomenon. When untreated, this condition can result in RV overload and congestive heart failure compromising patient survival. Repeated cardiac surgeries have been associated with greater morbidity and worse clinical outcomes, making transcatheter closure techniques an attractive treatment modality.

2 | CASE REPORT

A 22-year-old patient with complex congenital heart disease and multiple cardiac surgeries was referred to our center for rapidly progressive dyspnea with class III NYHA congestive right heart failure symptoms and hemolysis. She had complete repair of type B interrupted aortic arch in the neonatal period and aortic valve commissurotomy for severe aortic bicuspid valve stenosis at 11 years of age. Konno procedure and aortic valve replacement (23 mm St. Jude Medical valve) were performed for recurrent aortic stenosis at 19 years of age. She was largely asymptomatic upon regular follow-up with normal cardiac function. Upon admission, physical examination showed continuous murmur along the left parasternal region. The brain natriuretic peptide levels were elevated at 441 pg/ml. Color-flow Doppler transthoracic echocardiography detected an abnormal turbulent high-velocity ($V_{\text{max}} = 5 \text{ m/s}$) jet-flow from the aortic root to the RV, suggesting a possible fistula. The RV was dilated with pressure overload and moderately elevated mean arterial pressure at 30 mmHg. The mechanical aortic valve had normal function. Full infective endocarditis workup came negative. The patient was treated with diuretics. Computed tomography angiography (CTA) and transoesophageal echocardiography (TOE) delineated a 4–5 mm wide tunnel from the aortic root into the RV (Figure 1A,B). The coronary arteries were not involved. Appropriate informed consent was obtained for attempted device closure. The procedure was performed under general anesthesia, CTA-fluoroscopy fusion control, and TOE guidance. The left femoral arterial access was obtained using a 5F short introducer. Intravenous cefazolin was administrated. Heparin was not given as the patient was therapeutic on Fluindione. The left-to-right shunt was confirmed (QP:QS = 1.6:1 with PVR/SVR = 0.1). Contrast hand-angiography of the ascending aorta delineated the tunnel that was easily crossed using a 4F Judkins right coronary catheter (Cordis Corporation) in combination with an angled 0.035 in × 260 cm Radifocus Hydrophilic Guidewire M (Terumo Corp.). The wire was replaced with a stiff Type, angled 0.035” Radifocus Guidewire M (Terumo Corp.) positioned in the pulmonary arteries. Over the wire, a standard 7F Launch coronary guiding catheter (Medtronic, Inc.) was advanced and positioned in the RV. A 6 × 4 mm Amplatzer Duct Occluder II (ADO II) device (AGA Medical) was then delivered and deployed into position under fluoroscopic and TOE guidance (Figure 2). The device was released after confirmation of the absence of residual shunt by hand injection and the absence of interference...
with aortic valvular function on TOE (Figure 3A). The coronary circulation was not compromised on control angiography. Control ultrasound after 48 h showed good position of the device without residual shunt (Figure 3B). Diuretics were stopped after 2 weeks with concomitant resolution of hemolysis. Six months follow-up confirmed the absence of complications and full recovery (Figure 3C).

### DISCUSSION

We report a rare case of an aorta-to-RV fistula that was diagnosed in a young patient using multimodality imaging and efficiently treated with retrograde implantation of ADO II. Aorta-to-RV fistula has been most commonly seen after rupture of sinus of Valsalva aneurysm,
trauma, or as a complication of infective endocarditis and aortic dissections.\textsuperscript{1–3} These lesions have been very rarely described following transcatheter or surgical aortic valve replacements and ventricular septal defect repairs.\textsuperscript{4–8} Postoperative pericardial adhesions may prevent free aortic rupture into the pericardial space, increasing the predisposition to penetrate neighboring cardiac cavities, such as the RV. There are reports of suture failure associated with infective endocarditis but this hypothesis was ruled out in our case.\textsuperscript{5} It could also be related to iatrogenic injury/erosion from the displacement of heavily calcified tissue, the use of larger prostheses with the need for aggressive surgical debridement of the annulus, and depth of prosthesis implantation.\textsuperscript{2,4–6} The exact mechanism of fistula formation remains unclear. In our case, it is postulated to be secondary to suture failure with secondary progressive surgical patch erosion as detailed in Figure 1C.

We illustrate the diagnosis and treatment of this rare complication through multimodality fusion imaging. Although aortography is the gold standard for diagnosis, noninvasive methods such as CTA, TOE, and MRI are currently preferred and can be very helpful for the planning, the device choice, and during the procedure.\textsuperscript{2,3} Surgery is the primary treatment option; however, transcatheter closure has been attempted in a reasonable number of cases with encouraging short and midterm results. Our patient was considered a high-risk surgical candidate with a history of repeated cardio-thoracic surgeries making the percutaneous approach an appealing treatment option. The devices used for similar interventions include coils, and various devices from the Amplatzer family.\textsuperscript{1,7,8} Based on the literature review and our own experience, ADO II was the most suitable device. The device was perfectly fitted across the tunnel achieving complete shunt closure and patient clinical recovery. To our knowledge, this is the second report of aorta-to-RV fistula retrograde closure.\textsuperscript{7} The tunnel was easily accessed from the aorta using proper wires and the ADO II device was promoted directly through the RV to the fistula and the aorta with three-dimensional multimodality fusion imaging. The retrograde delivery avoided technical complications of arterio-venous circuit shortening procedure time and reducing irradiation. In conclusion, we believe that through this report, we strengthen the potential use of transcatheter closure techniques, even in the very rare cases of aorta-to-RV fistula. The anatomic complexity and morphological variability of these anomalies make them sometimes challenging to close using devices not originally designed for this purpose.

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CONFLICT OF INTERESTS
The authors declare that there are no conflict of interests.

ETHICS AND INFORMED CONSENT
The authors assert that all procedures contributing to this study comply with the ethical standards of the relevant national guidelines on human experimentation, and with the Helsinki Declaration of 1975, as revised in 2008. The patient’s signed informed consent was obtained for the reported procedures.

AUTHOR CONTRIBUTIONS
Raymond N. Haddad collected all clinical data, designed all illustrative material, and took the lead in writing the entire manuscript. All authors have revised and approved the final version of the manuscript.
REFERENCES


