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CASE REPORT

ADVANCED

CLINICAL CASE

# Cardiac Arrest Secondary to Traumatic Aortopulmonary Window During Transcatheter Pulmonary Valve Implantation in Supported Ross

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## ABSTRACT

We present a patient with a supported Ross procedure and severe pulmonary homograft stenosis who developed cardiac arrest while undergoing transcatheter pulmonary valve replacement and was found to have a large iatrogenic aortopulmonary window. Cardiopulmonary resuscitation was initiated followed by covered stent placement, extracorporeal membrane oxygenation support, and ultimately emergent surgery with a good outcome. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2019;1:746–50) © 2019 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 23-year-old man with a history of severe bicuspid aortic stenosis requiring percutaneous balloon valvuloplasty followed by a supported Ross procedure using a 32-mm sinus of Valsalva graft and a 30-mm right ventricle-to-pulmonary artery (RV-PA) homograft in 2016 presented with new-onset exercise intolerance. Echocardiography demonstrated severe RV-PA conduit stenosis with a peak gradient of 86 mm Hg. Cardiac magnetic resonance imaging showed severe RV-PA homograft narrowing (**Figure 1**). He was referred to the catheterization laboratory for transcatheter pulmonary valve replacement (TPVR).

## MEDICAL HISTORY

There was moderate to severe narrowing of the RV-PA homograft on angiography. Serial balloon dilation to 22 mm using ultra-high-pressure balloons was performed. Aortography with balloon dilation showed good flow to the coronary arteries. Following 22-mm balloon dilation, an internal intimal peel of the mid homograft was observed, with a small contained anterior contrast extravasation (**Figure 2**). Given the small injury, a 50-mm Palmaz bare-metal stent was chosen. On the basis of our inventory and the balloon length required, for a 30-mm homograft, a 26 × 6 BIB catheter (B. Braun Inc., Bethlehem, Pennsylvania) was selected, which was inflated at low pressure in the right ventricular outflow tract (RVOT). However, immediately following stent deployment, the patient became profoundly hypotensive with ventricularization of the aortic tracing.

## DIFFERENTIAL DIAGNOSIS

The differential diagnosis included coronary artery compression, acute conduit rupture, and acute aortic insufficiency from distortion of the neoaorta.

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Informed consent was obtained for this case.

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## LEARNING OBJECTIVES

- Serious complications are rare but can occur during TPVR.
- Identification of suitable candidates for TPVR is critical, and special precautions should be taken in patients with prior procedures, particularly Ross and arterial switch procedures.
- Differential diagnoses for hemodynamic instability during TPVR should include mechanical complications in addition to coronary artery compression.
- Although TPVR is widely performed and considered relatively safe, it should be performed only at centers of excellence where congenital cardiac surgical backup is available, as recommended by the 2018 AHA/ACC adult congenital heart disease guideline.

## INVESTIGATIONS

Given primary concerns for coronary artery compression, aortic root injection was performed. This showed a large traumatic aortopulmonary (AP) window posteriorly at the upper end of the stent with free flow of contrast from the aorta to the pulmonary artery (**Figure 3**, Video 1).

## MANAGEMENT

The patient further deteriorated and went into pulseless electrical activity arrest requiring cardiopulmonary resuscitation for 6 min. After return of spontaneous circulation, the decision was made to implant a covered stent at the area of the AP window to stabilize the patient. A 26 × 5 mm covered CP stent (B. Braun Inc., Bethlehem, Pennsylvania) was successfully implanted. Despite appropriate stent position confirmed by fluoroscopy, the patient decompensated and went into ventricular fibrillation, for which he received cardiopulmonary resuscitation for another 9 min. Percutaneous venoarterial extracorporeal membrane oxygenation support was initiated during cardiopulmonary resuscitation, and the arrhythmia was terminated when full support was achieved. After end-organ perfusion was ascertained, the patient was emergently taken to the operating room for surgical repair. A careful examination in the operating room revealed a largely contained tear in the aorta at the superior anastomosis of the supported Ross immediately adjacent to the superior RV-PA homograft suture line (**Figure 4**). He ultimately underwent repair of the ascending aorta using a 28-mm interposition graft and RV-PA conduit replacement using a 30-mm homograft. There were no post-operative complications. The patient had a full neurological recovery and was discharged on post-operative day 4.

## DISCUSSION

The Ross procedure is a widely accepted procedure for the management of aortic valve disease in children and young adults. This procedure leaves a patient with a prosthetic valve in the pulmonary position and a pulmonary autograft in the aortic position (neoaortic valve); both require long-term monitoring for subsequent reintervention.

Since its approval, TPVR has been widely performed as an alternative to surgery in selected patients with RVOT stenosis or insufficiency. Prospective TPVR trials have demonstrated satisfactory short- and mid-term outcomes with excellent post-intervention hemodynamic status (1). The most common complications are conduit injury, particularly with homografts, and stent fracture leading to RVOT reintervention. All implanting centers are encouraged to have covered stents available in case of significant conduit injury, but not all conduit injuries require covered stents. Serious procedural adverse events are rare. Coronary artery compression occurs in about 5% and is detected with aortography during balloon inflation, resulting in procedural termination. The current practice guidelines recommend performing ascending aortography and/or selective coronary angiography during the procedure to avoid such complication. Mechanical complications such as RVOT rupture occur less frequently but require urgent resuscitation and time-sensitive management decisions to avoid catastrophic outcomes.

There have been a number of previous reports on the occurrence of AP fistulae following TPVR in Ross (2) and non-Ross (3) patients, however, AP windows in previous reports were generally smaller and remained undetected until patients developed heart failure during follow-up.

We surmise that this supported Ross patient had scars along the suture lines in both the ascending aorta and main pulmonary artery that coalesced and healed as a unit, rendering a weak point that was prone to dehiscence. Inspection of the aorta during open repair in this patient confirmed that the AP window occurred in this region. Similarly, patients with transposition of the great arteries who underwent the arterial switch operation are also at increased risk for AP window after percutaneous pulmonary artery intervention because of the close

## ABBREVIATIONS AND ACRONYMS

**AP** = aortopulmonary

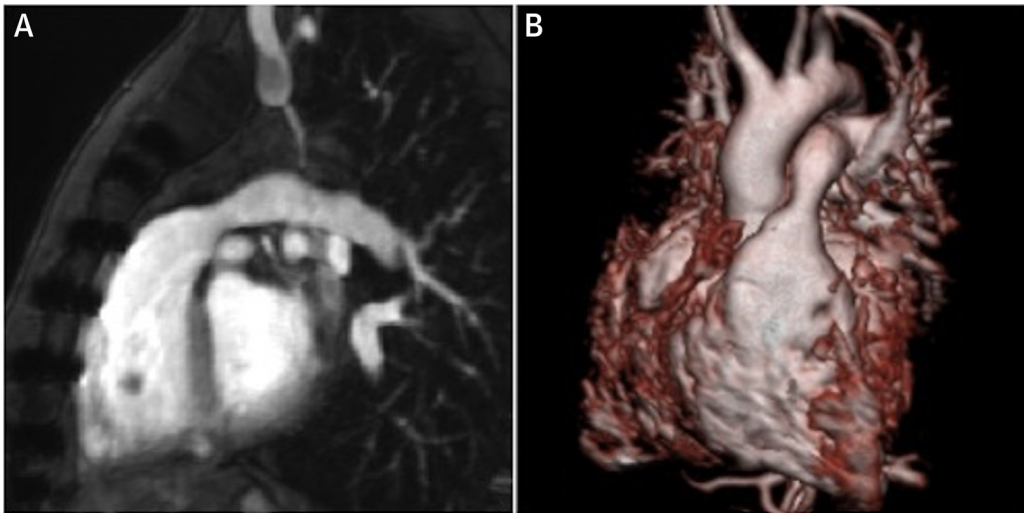
**RVOT** = right ventricular outflow tract

**RV-PA** = right ventricle-to-pulmonary artery

**TPVR** = transcatheter pulmonary valve replacement

JACC: CASE REPORTS, VOL. 1, NO. 5, 2019
DECEMBER 18, 2019:746–50

**FIGURE 1 Pre-Procedural Imaging Studies**

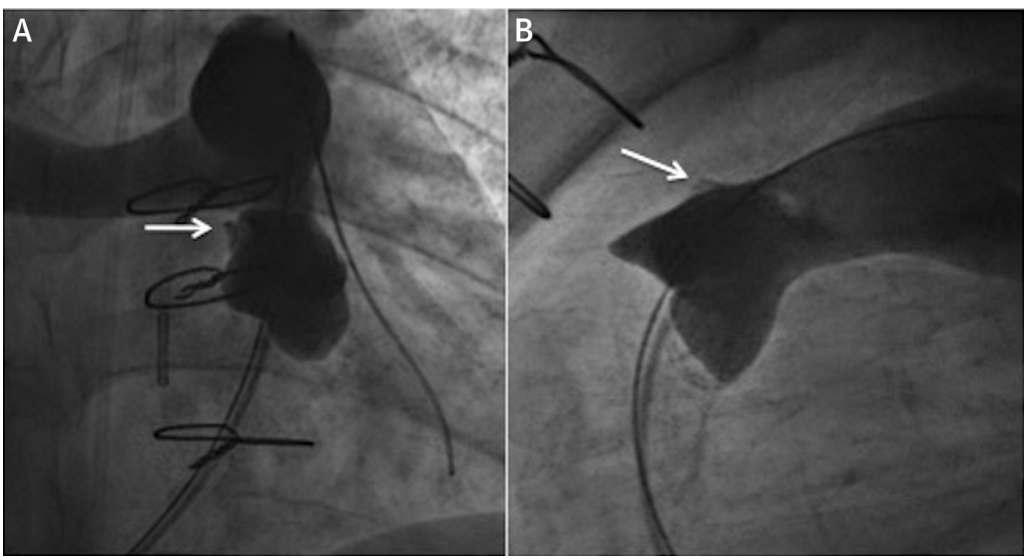



Severe homograft stenosis as demonstrated by cardiac magnetic resonance imaging in **(A)** right ventricular outflow tract (RVOT) sagittal and **(B)** 3-dimensional reconstruction. The relationship between the aorta and the RVOT is illustrated.

proximity of the great vessels as a result of the Lecompte maneuver (3,4).

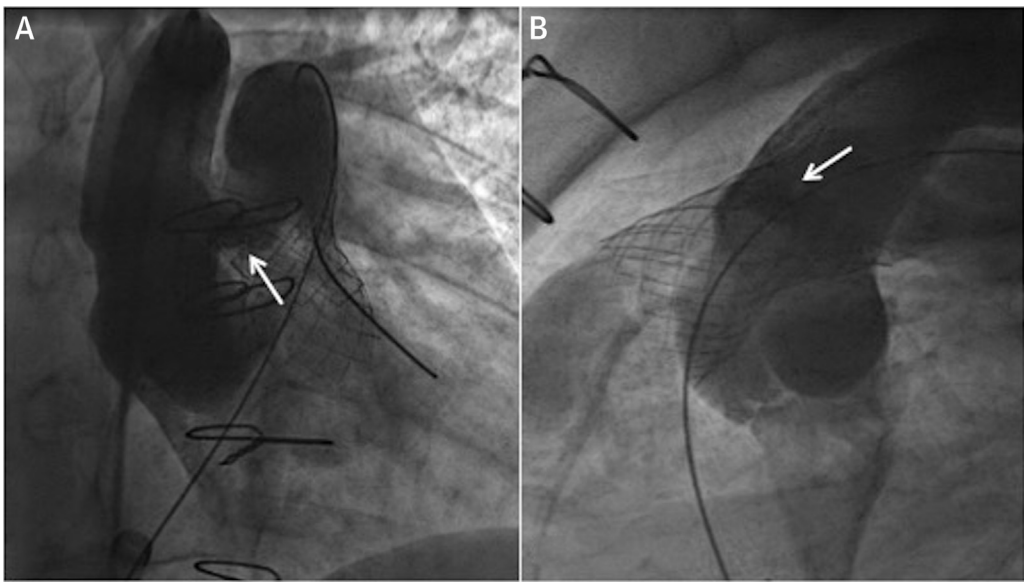
Cardiac magnetic resonance imaging is commonly performed for pre-procedural planning in TPVR. It provides information on RVOT and pulmonary artery size, geometry, and coronary anatomy, important for determining procedural suitability. Nonetheless, the risk for vessel injury and/or rupture on the basis of imaging data has not been described, and there is currently no consensus on good imaging modalities to predict such risk. Our case emphasizes that despite meeting criteria for appropriate patient selection,

**FIGURE 2 Right Ventricular Outflow Tract Angiography Following Balloon Dilation**




**Arrow** indicates mild contained conduit injury in **(A)** anteroposterior and **(B)** lateral projections.

**FIGURE 3** Ascending Aortography Following Right Ventricular Outflow Tract Stent Implantation



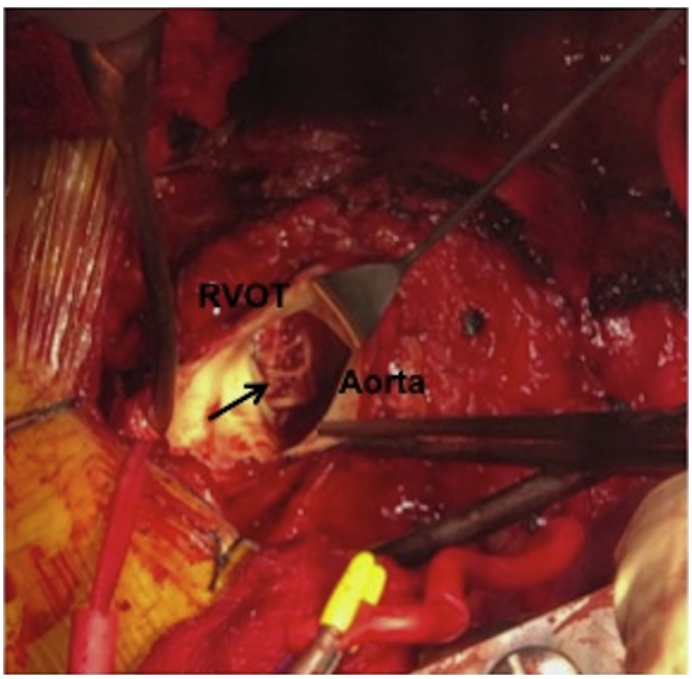

There is a large aortopulmonary window **(arrow)** at the upper end of the supported Ross graft with extravasation of contrast into the pulmonary artery in **(A)** anteroposterior and **(B)** lateral projections. See Video 1.

there is a risk for adverse events that is not always predictable.

The rupture may be attributed to the type of stent or size of balloon used. We debated the use of a covered stent initially but believed that the bare-metal stent would suffice given the small injury. Because of limitations in balloon length availability, a 26-mm BIB catheter was chosen for stent implantation in this 30-mm homograft. In our experience, stents usually measure smaller than the rated size when these balloons are used. In retrospect, it was unlikely that the covered stent would have reached the location of the AP window, as it was far superior, and this may have occurred regardless upon subsequent balloon inflation during valve implantation.

Management of iatrogenic AP windows was previously described (2,3). Covered stents and occluder devices have shown good results in hemodynamically stable patients with relatively small shunts. In patients with hemodynamic instability, emergent surgical repair is indicated, and temporary balloon occlusion or covered stent placement to attempt initial stabilization pending corrective surgery may be considered. Prompt response and the ability to immediately establish good hemodynamic support remain key in patients in unstable condition.

**FIGURE 4** Open Surgical Repair




Aortopulmonary window is demonstrated **(arrow)** with direct visualization of the right ventricular outflow tract (RVOT) stent from the aorta.

JACC: CASE REPORTS, VOL. 1, NO. 5, 2019
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## FOLLOW-UP

The patient was followed in the adult congenital heart disease program, with good recovery and normal valve and biventricular function.

## CONCLUSIONS

Serious complications following TPVR can occur despite careful patient selection and planning. Such cases should be carefully considered and should be undertaken only with the appropriate expertise, congenital cardiac surgical backup, and emergency equipment available to prevent a detrimental outcome (5).

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**KEY WORDS** complications, endovascular procedures, fistula, heart valve diseases, heart valve prosthesis implantation, pulmonary valve

**APPENDIX** For a supplemental video, please see the online version of this paper.