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CASE REPORT

CLINICAL CASE

Percutaneous Closure of an Iatrogenic Intracardiac Shunt in Treating Torrential Tricuspid Regurgitation

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ABSTRACT

A patient presented with severe right heart failure due to a large LV-to-RA shunt with left-to-right shunting and torrential tricuspid regurgitation 6-weeks following surgical sub-aortic stenosis resection. Retrograde delivery of an Occlutech ventricular septal defect device produced instantaneous resolution of shunt, reduction in tricuspid regurgitation, and impressive diuresis of 28 kg. (J Am Coll Cardiol Case Rep 2024;29:102389) © 2024 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 34-year-old woman with symptomatic severe congenital fibromuscular subaortic stenosis and a Doppler-derived systolic gradient of 130 mm Hg underwent surgical resection, complicated by postoperative complete heart block requiring dual-chamber pacemaker implantation and an iatrogenic left ventricular (LV) to right atrial (RA) shunt. The shunt was present but reported small on predischarge transthoracic echocardiography (TTE), with normal right ventricular (RV) size and function and moderate tricuspid regurgitation (TR); therefore, it was managed medically. The patient re-presented 6 weeks postoperatively with severe congestive heart failure and profound peripheral oedema. Her pulse was 102 beats/min, blood pressure was 102/61 mm Hg, and jugular venous pulse was visible at +7 cm. Thoracic auscultation was consistent with a right pleural effusion, there was gross peripheral oedema to the midabdomen but no pulsatile hepatomegaly. Her admitting weight was 118 kg, and she had gained 7 kg despite 240mg intravenous furosemide infusion.

LEARNING OBJECTIVES

- To be able to make a differential diagnosis of right ventricular volume overload following surgical resection of severe fibromuscular subaortic stenosis.
- To understand the role of percutaneous closure of a large left-to-right intracardiac shunt in reducing right ventricular volume overload and relieving torrential tricuspid regurgitation.

PAST MEDICAL HISTORY

There was nothing else of note in the patient's medical history.

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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.

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ABBREVIATIONS AND ACRONYMS

3D-VCA = 3-dimensional vena contracta area

LA = left atrium

LV = left ventricular

PAP = pulmonary arterial pressure

RA = right atrium

RV = right ventricle

TEE = transesophageal echocardiogram

TR = tricuspid regurgitation

TTE = transthoracic echocardiogram

TV = tricuspid valve

VSD = ventricular septal defect

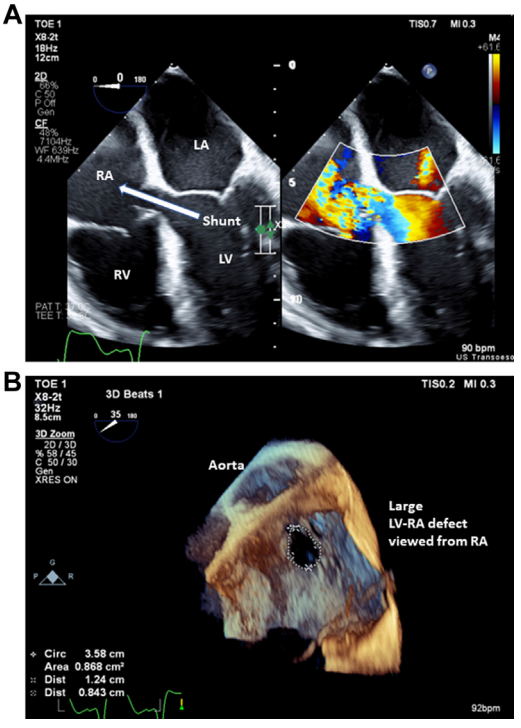
DIFFERENTIAL DIAGNOSIS

Differential diagnosis included recurrent subvalvular stenosis, worsening LV-RA shunting, worsening TR, and infective endocarditis.

INVESTIGATIONS

Chest radiography revealed a small, right-sided pleural effusion. TTE revealed normal LV size with an ejection fraction of 65%, dilated RV (end diastolic area = 11.2 cm^2/m^2), dilated RA of 5.3 cm, RV forward stroke-volume of 77 mL, and pulmonary arterial pressure (PAP) at 38 mm Hg. A transesophageal echocardiogram (TEE) demonstrated residual subaortic stenosis (peak gradient = 13 mm Hg), a large LV-RA shunt (**Figure 1**) measuring 12 × 9 mm situated immediately above the septal leaflet of the tricuspid valve (TV) (Video 1) causing significant left-to-right shunt (QP:QS = 1.6:1) (Video 2), and torrential central tricuspid regurgitation (TR) (**Figure 2**, Video 3) with a 3-dimensional vena contracta area (3D-VCA) measuring 1.58 cm^2 owing to failure of TV leaflet coaptation (**Figure 3**). The RV pacing wire was noted adjacent to the posterior aspect of the TV septal leaflet toward the posteroseptal commissure but was not impinging TV leaflet motion.

FIGURE 1 Large LV-RA Defect on 2D and 3D Echocardiography



(A) Four-chamber and (B) 3-dimensional en face large LV-RA defect (12 × 8 mm; area = 0.89 cm^2), with significant LV-RA flow on color Doppler imaging.

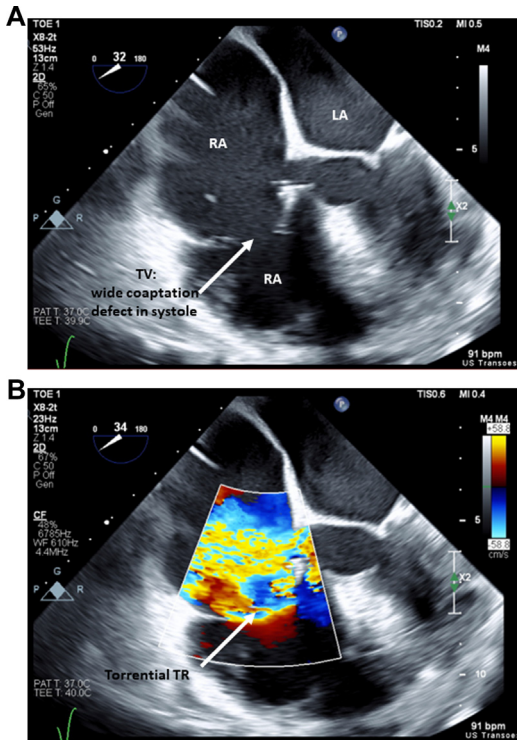
MANAGEMENT

A multidisciplinary team discussion suggested percutaneous closure of the LV-RA shunt with prior aggressive medical treatment of heart failure. The maximum tolerated intravenous furosemide was 360 mg/24 hours owing to hypokalemia (metolazone 1.25 mg not tolerated), and oral amiloride (5 mg) and spironolactone (50 mg) were commenced. At procedure, right femoral vein (8-F) and right femoral arterial (5-F) access was achieved, but it proved impossible to cross the LV-RA defect antegrade from the RA and achieve a stable catheter and wire position owing to the torrential left-right shunt (Video 4). A retrograde arterial catheter and wire were therefore passed from LV to RA to inferior vena cava and snared out of the right femoral vein (Video 5). A retrograde arterial 8-F long delivery sheath was then passed LV to RA, being careful not to trap the pacing lead. Sizing of closure device was based on TEE assessment of the defect dimensions. A 14-mm Occlutech muscular ventricular septal defect (VSD) occluder was delivered successfully across the LV-RA shunt (Videos 6 to 8), with instantaneous increase in blood pressure (from 99/54 mm Hg to 156/86 mm Hg) and reduction in heart rate (from 156 to 77 beats/min). Intraprocedural TEE demonstrated no residual shunt across the VSD (Video 9) and an immediate reduction in TR severity (Video 10) from torrential to severe (3D-VCA 1.58 cm^2 to 0.72 cm^2) (**Figure 3**). Intravenous diuretics were continued for 4 days before being converted to oral diuretics. The patient clinically improved significantly following a rapid diuresis, and she was discharged 7-days post-procedure, having lost a total of 28 kg.

DISCUSSION

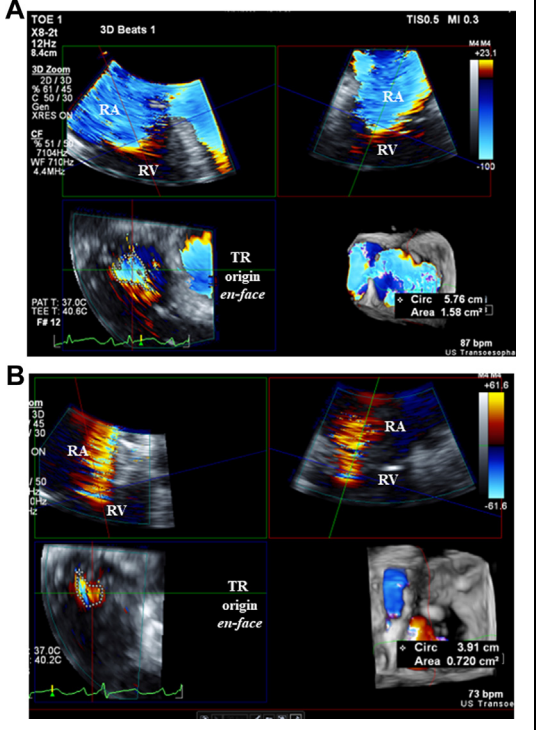
Left ventricular to right atrial (LV-RA) shunts, first reported by Gerbode et al in 1958[1], are ventricular septal defects that can be congenital (26.4%) or acquired (72.7%) in origin.[2,3] The ventricular septum that forms part of the subaortic outflow tract is closely adjacent to the membranous atrioventricular septum and septal attachment of the TV. Excision of the fibromuscular shelf of subaortic stenosis may disrupt the conduction axis (resulting in complete heart

FIGURE 2 Torrential Tricuspid Regurgitation



(A) Two-dimensional failure of TV leaflet coaptation in systole (B) with torrential TR.

FIGURE 3 Reduction of Tricuspid Regurgitation Following Closure of LV-RA Defect



(A)Preprocedural torrential TR (3D vena contracta area = 1.58 cm^2). (B) Immediately after successful VSD occluder device implantation (3D vena contracta area = 0.72 cm^2).

block), the atrioventricular septum, and TV attachment (particularly in patients with a large membranous septum), so that blood traverses from the LV, through the membranous atrioventricular septum, to the RA above the TV. The large systolic pressure gradient between LV and RA results in a high-velocity systolic flow from LV to RA, which can be difficult to diagnose on TTE before RV dilatation, because the LV-RA shunt is often mistaken for a TR jet and missed in up to 38% of cases.[4] Increasing volume loading of the right heart can lead to increasing TR of an uncommon etiology[5] and precipitation of right heart failure. In our patient's case, the LV-RA shunt was repaired perioperatively with interrupted sutures so that only a small LV-RA shunt was reported on discharge TTE. The presumption was the dehiscence of the sutures a short while post-discharge, resulting in a much larger LV-RA defect with increasingly severe symptoms.

Transcatheter closure of an LV-RA defect using an Amplatzer occluder device has been used to treat various left-to-right intracardiac shunts[6] and acquired defects after surgical VSD closure.[7] In our case, transcatheter closure of an acquired VSD following surgical subaortic stenosis resection had the advantage of avoiding the inherent risks of repeated cardiopulmonary bypass, and our patient already had a dual-chamber pacemaker owing to postoperative complete heart block. We chose to implant a muscular VSD because of the large size and relatively broad "neck" of the defect, in preference to other potential device choices (Amplatzer duct occluders, atrial septal defect occluders, VSD coils). Percutaneous implantation of a VSD occluder device can be performed antegrade or retrograde depending on operator choice; in our patient, an antegrade approach proved impossible because of shunt turbulence, while retrograde approach was successful. Deployment of an Occlutech occluder device across the LV-RA defect produced dual immediate intraprocedural hemodynamic effects: obliteration of the large left-to-right shunt and instantaneous significant reduction in TR severity. Both were associated with

rapid normalization of the patient's blood pressure and permitted substantial off-loading of the right-heart to enable rapid diuresis and early reverse RV remodeling.

FOLLOW-UP

At 3-months post-discharge, the patient was in NYHA functional class I. TTE reported no residual LV-RA shunt, significant RV reverse remodeling (end-diastolic area = 8.72 cm^2/m^2), only mild TR, and PAP 27 mm Hg.

CONCLUSION

Extensive surgical resection of severe fibromuscular subaortic stenosis may disrupt the atrioventricular septum, resulting in an acquired LV-RA shunt, which if large can cause significant RV overload and torrential TR. Antegrade transcatheter closure of the LV-RA defect may be difficult because of the high-velocity systolic jet. Retrograde arterial transcatheter closure is possible and may completely obliterate left-to-right shunting with immediate reduction in TR severity, permitting significant RV off-loading, rapid diuresis, and early reverse RV remodeling.

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The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS left ventricle to right atrial shunt, tricuspid regurgitation, transcatheter ventricular septal defect closure

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