# Iatrogenic Aorta-to-right atrial fistula: a rare cause of pulmonary hypertension

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## Introduction

Aorta-to-right atrial fistula is an extremely rare late complication of prosthetic aortic valve replacement. Increased survival and improved patient outcomes following complex aortic valve surgeries poses paramount significance to the recognition of this unusual complication.

We describe the case of an 80-year-old woman who presented with slowly progressive symptoms of heart failure within a year of bioprosthetic aortic valve replacement. Initial transthoracic echocardiographic features were consistent with pulmonary hypertension, and later transesophageal echocardiography and cardiac magnetic resonance imaging revealed an aorta-to-right atrium fistula with shunt. The patient underwent successful revision surgery with aortic valve replacement, tricuspid valve repair and closure of fistula.

## Case presentation

An 80-year-old female presented with 2 month history of exertional dyspnea and reduced exercise tolerance. She was 10 months post aortic valve replacement with a pericardial aortic bioprosthesis, and a left internal mammary artery graft to left anterior descending artery, on a background of severe aortic stenosis and ischemic heart disease. Clinical examination demonstrated raised jugular venous pressure and a faint systolic murmur. NTproBNP was 935.

## Discussion

Aorto-cameral fistula refers to an abnormal vascular connection between the aortic root and one of the cardiac chambers. Aorta-to-right atrial fistula is a sub-type of this extracardiac connection. It may be congenital or acquired.

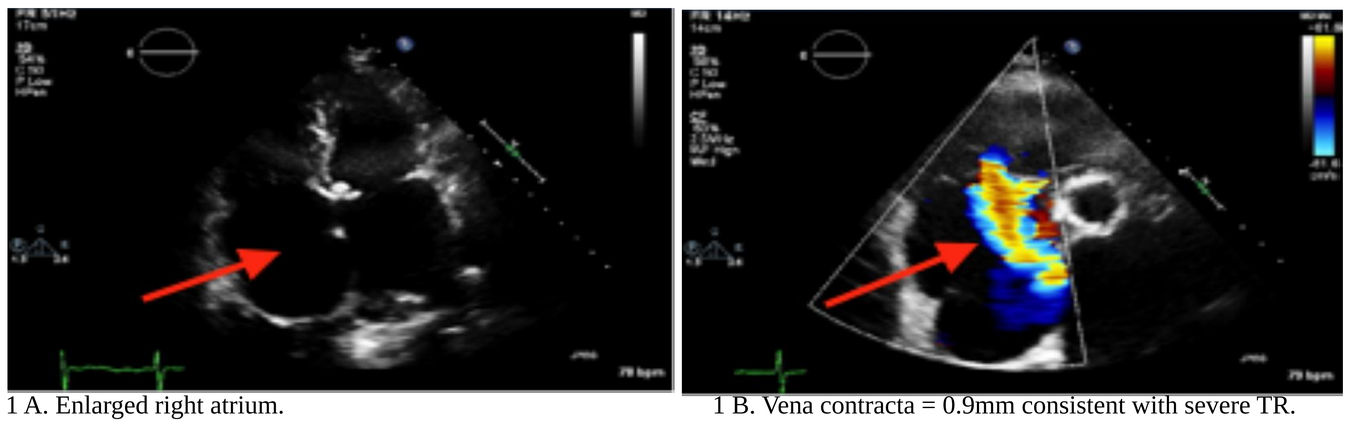
Most of the common acquired aetiologies of aorto-right atrial fistulas are related to its occurrence secondary to bacterial endocarditis, paravalvular abscess, aortic dissection, ruptured sinus of Valsalva, percutaneous closure of patent foramen ovale/atrial septal defect, or aortic valve surgery.

Several mechanisms may lead to the development of an aorta-right atrial fistula after aortic valve surgery including excessive and aggressive debridement of bulky area of calcification from the aortic annulus, iatrogenic injury to the interatrial septum, perivalvular damage attributable to improper retraction, oversized aortic prosthesis, infected tissue before valve replacement, and connective tissue abnormalities. All patients with new cardiac symptoms following an aortic valve surgery require assessment with echocardiography and/or computed tomography (CT) and CMR.

Management of this condition depends centrally on clinical presentation and the presence of pulmonary hypertension. Redo surgery is challenging and may be associated with high morbidity and mortality. Surgical intervention comprises of repairing the affected aortic segment, suturing of the fistula, replacing the prosthesis if the valve is destroyed and also correcting other anomalies that have occurred secondary to this complication.

## Investigations

**Transthoracic echocardiogram** revealed a dilated right atrium (RA) and right ventricle (RV), severe tricuspid regurgitation (TR), and a D-shaped septum in diastole suggesting raised RV end-diastolic pressure, possibly pulmonary hypertension (Figure:1).These findings were absent prior to cardiac surgery.



1 A. Enlarged right atrium.
1 B. Vena contracta = 0.9mm consistent with severe TR.
1 C. PSAX view showing D-shaped septum in Diastole.
1 D. Mal coaptation of tricuspid leaflets.

**Figure 1. Transthoracic echocardiography findings consistent with Pulmonary Hypertension.**

**Cardiac magnetic resonance imaging (CMR)** was performed to evaluate the unexplained TR. CMR raised the possibility of an aortic root to RA shunt, but aortic valve artefact limited complete evaluation. The RV to LV stroke volume ratio of 2:1 was more compatible with a shunt rather than isolated TR.
Subsequent **transesophageal echocardiogram (TEE)** colour Doppler demonstrated a systolic shunt through an echo-free space from the aortic root into the RA, converging with the severe TR jet, with evidence of RA and RV volume overload. (Figure:2)

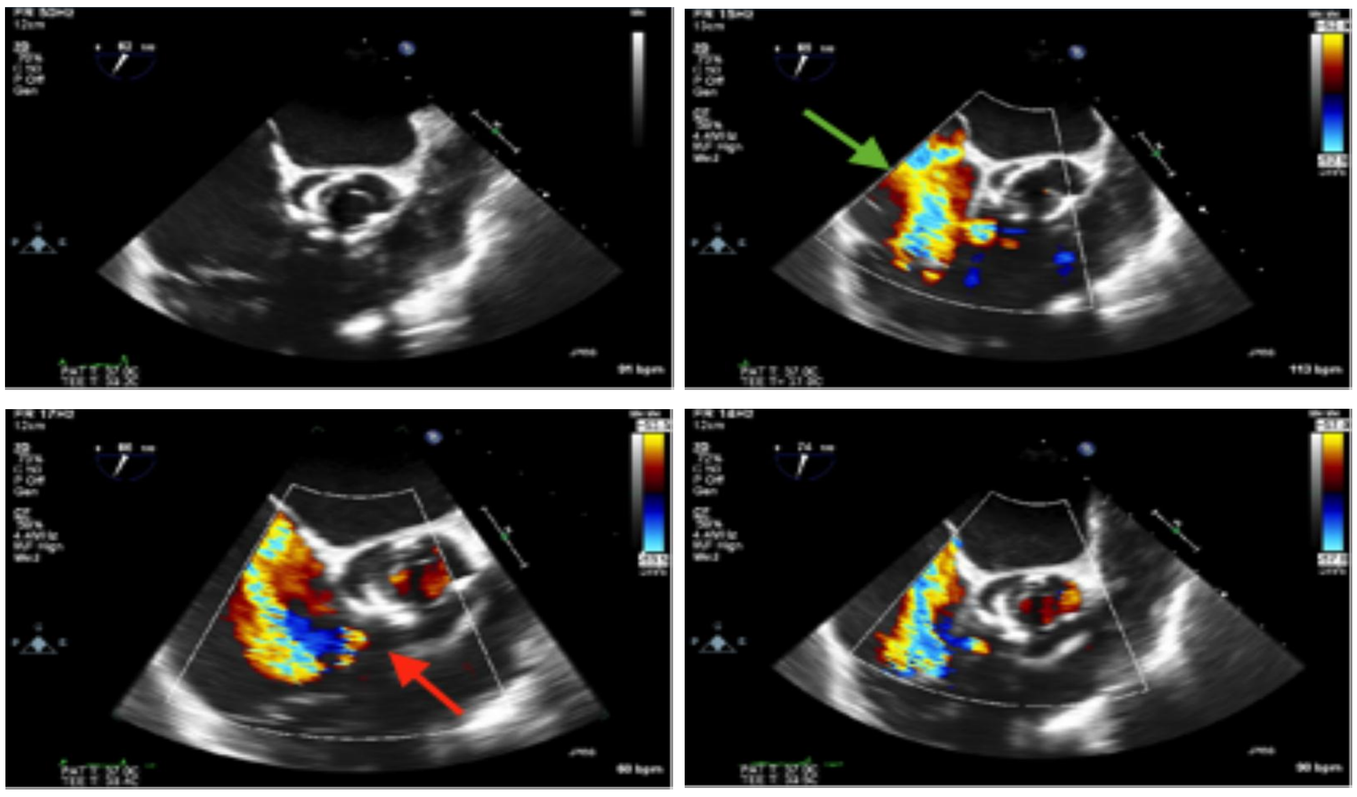


**Figure 2: Transoesophageal echocardiography demonstrating Severe TR and an Aorta-to-Right atrium fistula.**

## Treatment and Outcome:

The patient was referred back to the cardiothoracic surgeon in view of revision surgery. The prosthetic aortic valve was excised with placement of a new aortic bioprosthesis and suturing of the fistula, followed by insertion of a tricuspid annuloplasty band.
The patient made a good recovery without any complications and reported significant improvement in her symptoms in clinic follow-up at 8 weeks.

## Conclusion

Denovo cardiac symptoms and evidence of pulmonary hypertension on transthoracic echocardiogram in a patient post aortic valve surgery warrents further investigation with CMR and TEE, considering the possibility of aorta-to-RA fistula.