

# Pulmonary Artery Stump Thrombus in a Patient With Remote History of Rastelli Procedure



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## INTRODUCTION

The Rastelli procedure, which involves baffle closure of the ventricular septal defect (VSD) to the aorta, ligation of the pulmonary artery (PA) above the valve, and placement of a right ventricle (RV)-to-PA conduit, is a standard surgical technique for congenital dextro-transposition of the great arteries (D-TGA) with VSD and pulmonary stenosis.<sup>1</sup> The procedure creates a PA stump at the end that communicates with systemic circulation. Although prior literature has described that thrombi can form in ligated PA stumps in patients undergoing the Fontan operation in which thromboembolism can be attributed to low-flow state in the venous pathway, the risk for thromboembolism may be considered low in the PA stump connected to systemic circulation in clinical practice.<sup>2</sup> However, we present a patient with repaired D-TGA with Rastelli procedure and a possible long-term thrombotic complication.

## CASE PRESENTATION

A 37-year-old man with congenital D-TGA complicated with VSD and pulmonary stenosis, post Rastelli procedure at 6 years of age, with multiple revisions, stenting, and replacement of the RV outflow conduit throughout the patient's life, and heart failure with reduced left ventricular (LV) ejection fraction, presented to the emergency department with persistent palpitations. Upon arrival, the heart rate was measured at 105 beats/min with blood pressure of 112/78 mm Hg. Heart rhythm was regular, with early systolic murmur 2/6 auscultated at the left upper sternal border, and the lungs were clear to auscultation. The patient had prior known conduction abnormalities, including complete heart block necessitating dual-chamber permanent pacemaker implantation in their early 20s, which was subsequently upgraded to an implantable cardioverter-defibrillator after the patient was found to have inducible ventricular tachycardia during an electrophysiologic study at 32 years of age. Twelve-lead electrocardiography revealed atrial flutter with a rapid ventricular response, which was confirmed by implantable cardioverter-defibrillator interrogation. Transthoracic

echocardiography (TTE) was performed and confirmed significant LV dysfunction with D-TGA. The patient was started on apixaban and scheduled for direct cardioversion after burst pacing failed to terminate the tachyarrhythmia. Transesophageal echocardiography (TEE) was performed to rule out intracardiac thrombus.

TEE confirmed D-TGA, with aortic positioning anterior and rightward to the PA. The native PA stump was present, and the pulmonary valve (PV) was seen with opening leaflets. No thrombus was visualized in the left atrium and left atrial appendage, but there was an 8.5 × 12.8 mm elliptical mobile thrombus floating at the top of the PA stump (Figure 1, Video 1). Cardioversion was aborted because of concerns of dislodging the thrombus, which may have resulted in cardioembolic stroke. Apixaban was switched to intravenous heparin on the basis of its high efficacy in resolving LV thrombi from existing clinical evidence.<sup>3</sup> Serial targeted TTE was performed to monitor the thrombus, and 5 days later, TEE was repeated after the thrombus was no longer visualized on TTE, and it confirmed that no intracardiac thrombus was present (Figure 2, Video 2). The patient was subsequently electrically converted to normal sinus rhythm and discharged from the hospital without complications.

## DISCUSSION

D-TGA is a serious congenital cardiac anomaly with high mortality in the absence of surgical correction. Known for artificial redirection of ventricular outflows and reconstruction of the PA with artificial conduits, the Rastelli procedure used to be the preferred and conventional surgical approach for D-TGA.<sup>4</sup> The cardiac end of the native PA was oversewn as part of the procedure, leaving a PA stump. Despite significant improvement of early postoperative survival over the past decades, serious late complications have been reported, including LV dysfunction and RV outflow obstruction, which may potentially affect the life expectancy and quality of the recipients.<sup>5</sup> Arterial switch procedure using the LeCompte maneuver gradually became the main surgical approach because of reduction in reoperation rate.<sup>6</sup> But in patients with significant pulmonary stenosis and D-TGA who are not candidates for arterial switch, the Rastelli procedure is still performed, and patients who have undergone this operation can be seen in daily practice, so familiarity with its long-term complications remains crucial for clinicians. Thromboembolism at the PA stump has been rarely mentioned in prior literature as a potential complication, as the PA stump was usually sutured incorporating the PV during the Rastelli procedure to prevent thrombus from forming above the leaflet.<sup>7</sup> The presence of PV cusps should guide clinicians to pay more attention to the stump for possible thrombus formation. The PA stump can be overlooked on TEE in patients with atrial tachyarrhythmias, when the focus is typically on the left atrial appendage to exclude intracardiac thrombus. Clinicians who are preoccupied with atrial arrhythmia may not recognize the

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**VIDEO HIGHLIGHTS**

**Video 1:** Two-dimensional TEE, midesophageal biplane view (90° and 180°) of the PA stump, demonstrates a thrombus near the PA stump.

**Video 2:** Two-dimensional TEE, midesophageal 0° view of the PA stump after intravenous heparin anticoagulation, demonstrates the absence of intracardiac thrombus.

View the video content online at [www.cvcasejournal.com](http://www.cvcasejournal.com).

complexities of patients' congenital heart anomalies, and therefore, transfer to centers with adult congenital heart disease expertise may be warranted in such scenarios to provide patients with specialized care as needed.

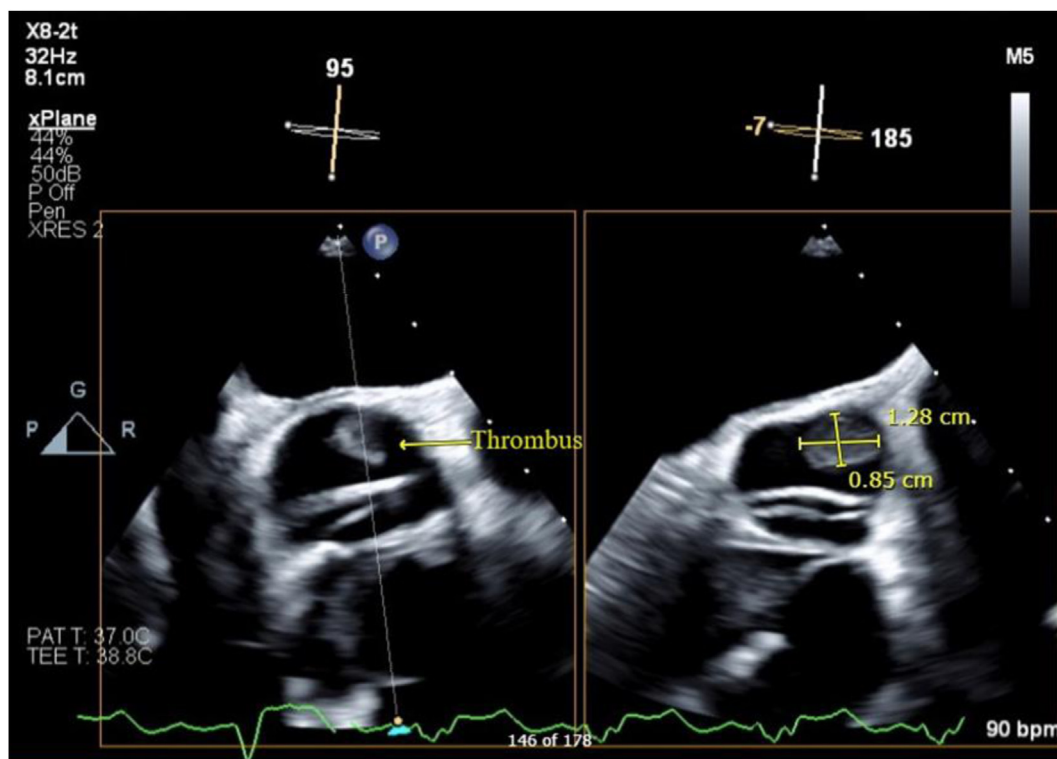
In addition to echocardiogram, cardiac computed tomography and cardiovascular magnetic resonance imaging have been proposed as complementary imaging modalities with growing potential. With its high spatial resolution and ability for three-dimensional reconstruction, cardiac computed tomography offers delineation of complex congenital cardiac anomalies and calcified structures such as homograft and prosthetic valves, while metallic artifacts from sternal wires and stents can often degrade cardiovascular magnetic resonance images.<sup>8</sup> However, the cumulative risk from radiation exposure in

cardiac computed tomography needs to be taken into consideration, especially as patients with congenital heart disease often seek care at early ages.<sup>9</sup>

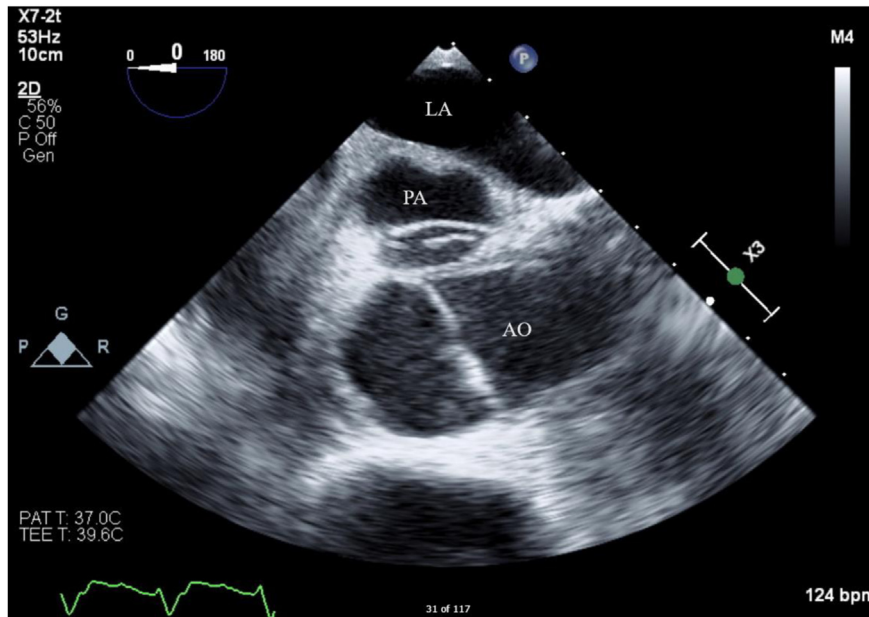
We describe a rare complication of the Rastelli procedure in which the PA stump can be a nidus for thrombus formation, especially in patients with LV dysfunction and atrial tachyarrhythmia. Thorough evaluation on echocardiography is essential in patients who have undergone the Rastelli procedure or other operations in which a PA stump is created to rule out intracardiac thrombus before cardioversion in an effort to reduce the complication of cardioembolic stroke. We propose that parasternal short-axis and long-axis views on TTE with individualized angle adjustment to visualize the pulmonic and aortic valve can be attempted first in Rastelli patients, followed by TEE if thromboembolic risks remain high or image quality is suboptimal. Understanding the concepts and potential complication profile of the Rastelli procedure remains essential, and expert consultation with congenital heart disease specialists is often appropriate in the care of these patients.

**CONCLUSION**

The presented case highlights the potential development of a thrombus within the PA stump in recipients of the Rastelli procedure for D-TGA. Vigilant and thorough echocardiographic assessment of the PA stump remains crucial to rule out thrombus before direct cardioversion.



**Figure 1** Two-dimensional TEE, midesophageal biplane view (90° and 180°) of the PA stump and aorta at end-diastole, demonstrates a thrombus near the PA stump.



**Figure 2** Two-dimensional TEE, midesophageal 0° view of the PA stump after heparin anticoagulation, demonstrates the absence of intracardiac thrombus. AO, Aorta; LA, left atrium.

### ETHICS STATEMENT

The authors declare that the work described has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans.

### CONSENT STATEMENT

Complete written informed consent was obtained from the patient for the publication of this study and accompanying images.

### FUNDING STATEMENT

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### DISCLOSURE STATEMENT

The authors report no conflict of interest.

### SUPPLEMENTARY DATA

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.case.2023.12.011>.

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