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CASE REPORT

INTERMEDIATE

CLINICAL CASE

# A Caseating Tricuspid Mass in an Adult With Eisenmenger Syndrome Secondary to Aorto-Pulmonary Window

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## ABSTRACT

We present the case of a 55-year old Caucasian man with Eisenmenger syndrome secondary to uncorrected aorto-pulmonary window, whose clinical course has been complicated by recurrent cerebral abscesses and dynamic tricuspid annular caseation with probable pulmonary embolization. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2023;8:101646) © 2023 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/).

## PAST MEDICAL HISTORY

The patient is a 55-year-old Caucasian man with Eisenmenger syndrome (ES) secondary to uncorrected aorto-pulmonary window (APW). His congenital heart disease was diagnosed in his late teens. He developed recurrent fungal cerebral abscesses in his twenties, which presented with focal seizures and required multiple neurosurgical procedures. He has residual scarring in the affected cerebral regions and continues to require multiple antiepileptic drugs for complex seizure disorder.

From a cardiac perspective, the patient had stable New York Heart Association functional class II symptoms in the initial decades following diagnosis with no hospitalizations for decompensation. Serial transthoracic echocardiograms (TTE) demonstrated a large APW (**Figures 1A and 1B,** Video 1), dilated right ventricle (RV) with marked hypertrophy and mildly impaired contractility, preserved left ventricular ejection fraction, mild-moderate pulmonary regurgitation, and progressively elevated pulmonary pressures (**Figures 2A to 2D**). He was commenced on Sildenafil in 2010, with a pretreatment right ventricle-right atrium (RV-RA) gradient of 106 mm Hg. Availability of multimodality cardiac imaging in recent years enabled further characterization of the patient's shunt, confirming a large (6.4-cm anteroposterior) APW formed by continuity of the distal main pulmonary artery with the mid-ascending aorta (**Figures 3A and 3B**).

### LEARNING OBJECTIVES

- To understand how markedly deranged physiology in adults with complex congenital heart disease can result in unique complications.
- To appreciate the role of serial multimodality cardiac imaging in the assessment of patients with complex congenital heart disease.

## HISTORY OF PRESENTATION

In 2017, the patient presented for routine cardiac review. He did not have any chest pain, worsening dyspnea, fevers, or constitutional symptoms. There was no history of intravenous drug use. Heart rate

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**ABBREVIATIONS AND ACRONYMS**

**APW** = aorto-pulmonary window

**CT** = computed tomography

**ES** = Eisenmenger syndrome

**RA** = right atrium

**RV** = right ventricle

**TTE** = transthoracic echocardiogram

was 72 beats/min and regular. Blood pressure was 118/70 mm Hg. There was peripheral cyanosis and clubbing of fingers and toes. Jugular venous pressure was elevated. Heart sounds were dual with prominent pulmonic component of the second heart sound. There was a grade 2/6 systolic murmur followed by a moderately long pulmonary regurgitation murmur. Chest was clear. There was no peripheral edema. There were no focal neurologic deficits nor any peripheral signs of embolization. Electrocardiogram demonstrated sinus rhythm with incomplete right bundle branch block, right axis deviation, and widespread T-wave inversion. Chest x-ray showed enlarged pulmonary trunk, central pulmonary plethora with peripheral pruning, and right-sided aortic arch (**Figure 4**). TTE demonstrated significant improvement in pulmonary pressures (RV-RA gradient 81 mm Hg) but revealed a new echodense calcific-rimmed mass on the tricuspid annulus (**Figure 5A,** Video 2).

## DIFFERENTIAL DIAGNOSIS

Differential diagnoses for the mass included annular calcification, vegetation, thrombus, and neoplasm.

## INVESTIGATIONS

The patient was admitted for further investigations given the new mass. Bloods revealed no elevation of inflammatory markers (white cell count 3.8 [4.0-10.0], C-reactive protein 1.6 [<5]) with normal renal function and bland urinalysis. Blood cultures and serum cryptococcal antigen testing were negative. Computed tomography (CT) cardiac chambers confirmed a large mass (2.1 × 1.9 cm in axial dimensions) along the inferior margin of the tricuspid annulus with a hypodense core and nonenhancing calcific border (**Figure 5B,** Video 3). There was no lymphadenopathy or pulmonary nodules.

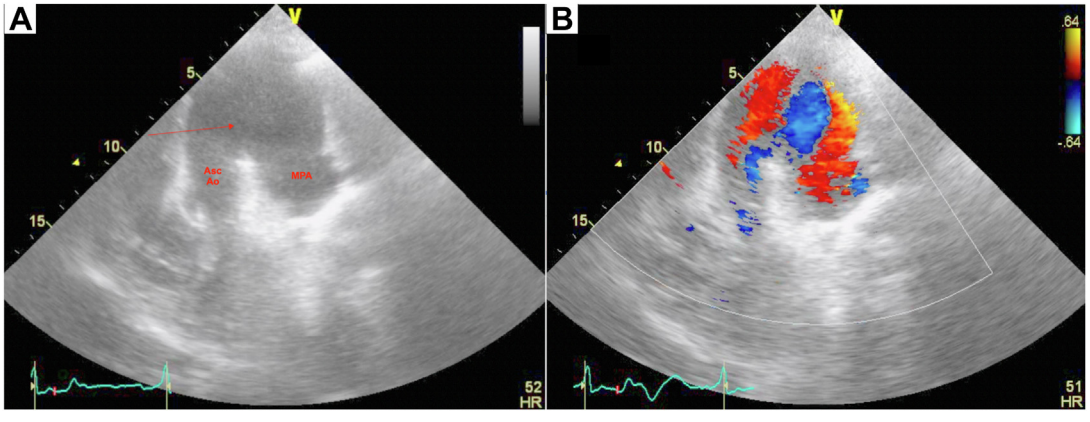
## MANAGEMENT

After review in a multidisciplinary team meeting involving cardiothoracic surgeons, radiologists, interventional cardiologists, and cardiac imaging specialists, a presumptive diagnosis of caseous tricuspid annular calcification was made with plans for serial imaging rather than invasive sampling.

Serial TTE and CT cardiac chambers in 2019 showed interval enlargement of the tricuspid mass (**Figures 5C and 5D,** Videos 4 and 5), now measuring 2.8 × 3.0 cm, with no Doppler evidence of obstruction. The patient's pulmonary hypertension continued to improve (RV-RA gradient 74 mm Hg) and he remained clinically well.

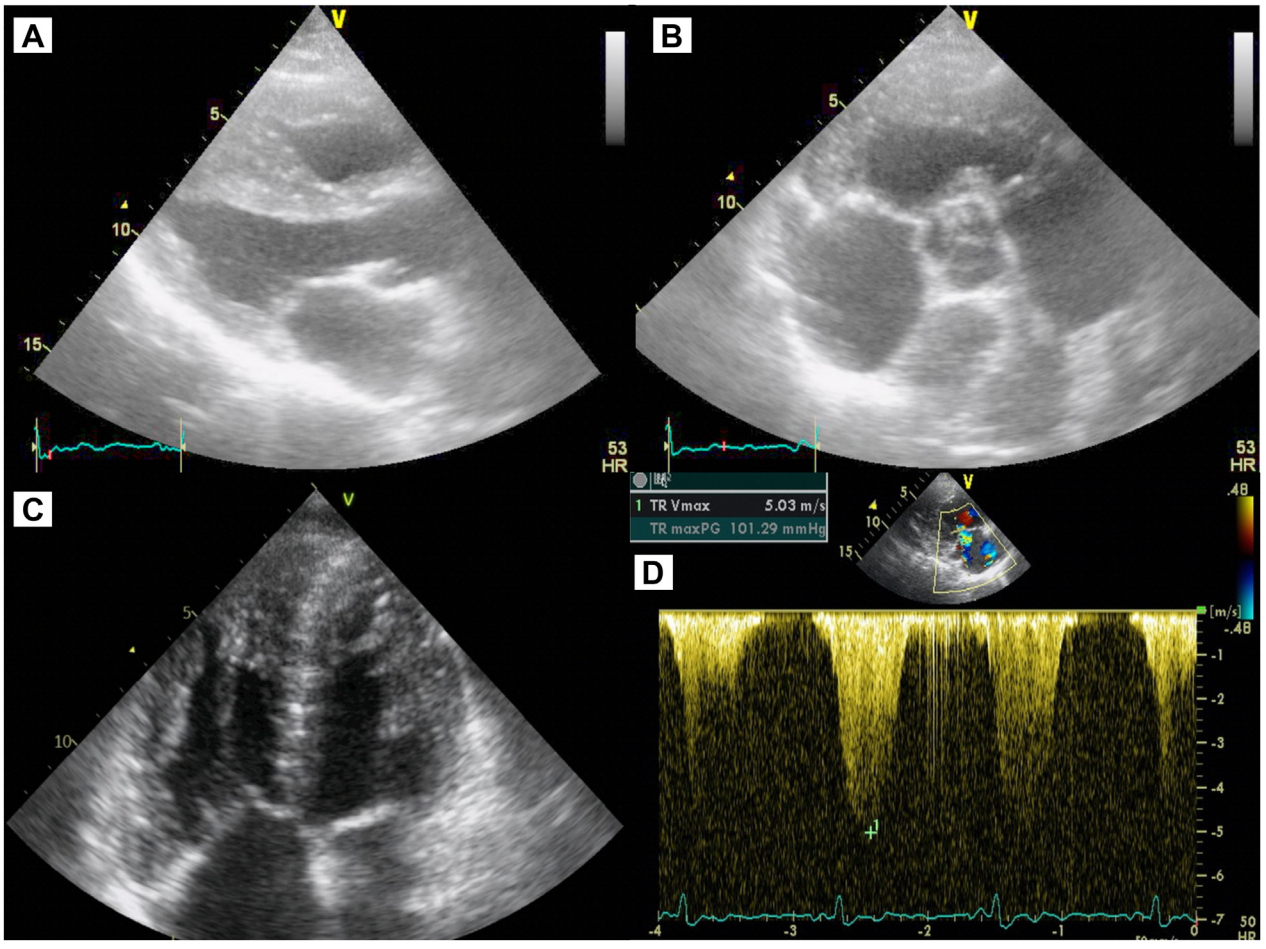
Repeat imaging in 2021 demonstrated a significant decrease in the size of the tricuspid mass, now measuring 2.2 × 1.9 cm (Videos 6 and 7). Of note, the central hypodense component of the mass had regressed and the calcified rim was more opposed (**Figures 5E and 5F**). There was concurrently a marked

**FIGURE 1 Transthoracic Echocardiogram Displaying Aorto-Pulmonary Shunt**




Suprasternal views on transthoracic echocardiogram demonstrating shunting across defect **(arrow)** between main pulmonary artery (MPA) and ascending aorta (Asc Ao).

**FIGURE 2** Pre-Treatment Transthoracic Echocardiogram




Pretreatment transthoracic echocardiogram demonstrating dilated and hypertrophied right ventricle **(A to C)** with severely elevated pulmonary pressures **(D)**.

rise in pulmonary pressure (RV-RA gradient 117 mm Hg), suggesting possible embolization of the mass to the pulmonary circulation (Figure 6). The patient now had worsening dyspnea and fatigue. CT pulmonary angiogram did not reveal discrete pulmonary emboli. He was subsequently hospitalized with rapid atrial flutter and intermittent runs of wide complex tachycardia. He underwent successful direct current cardioversion and was then commenced on amiodarone and warfarin. Repeat magnetic resonance imaging brain demonstrated stable regions of focal encephalomalacia but no new lesions to suggest cerebral embolization of the mass.

## FOLLOW-UP

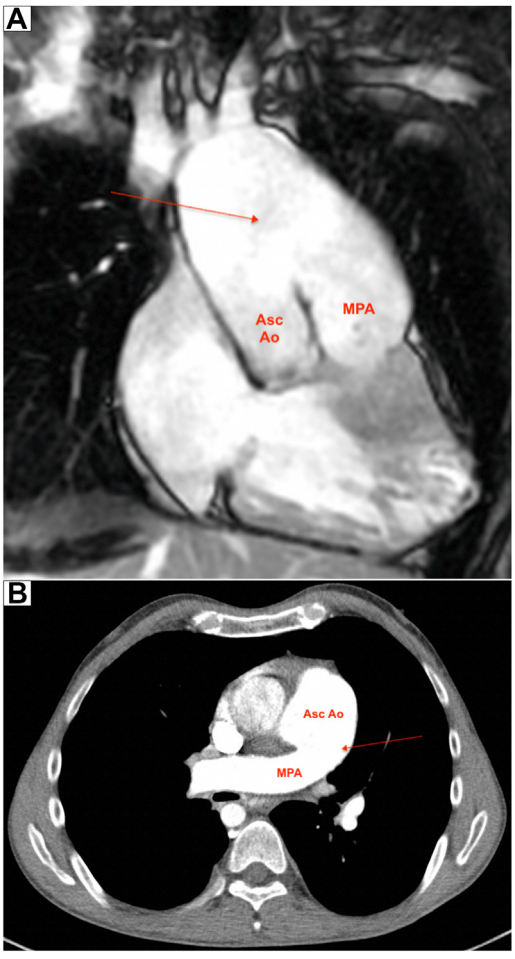
Given his progressive clinical decline, the patient has been referred for heart-lung transplant assessment.

## DISCUSSION

APW is an uncommon form of congenital heart disease in which abnormal septation of the truncus arteriosus produces a communication between the proximal aorta and main pulmonary artery.[1] Without early corrective surgery, severe pulmonary hypertension and ES develop in the first few months of life. Late presentation of APW in adult life is extremely rare and few surgically uncorrected cases survive into adulthood.[2]

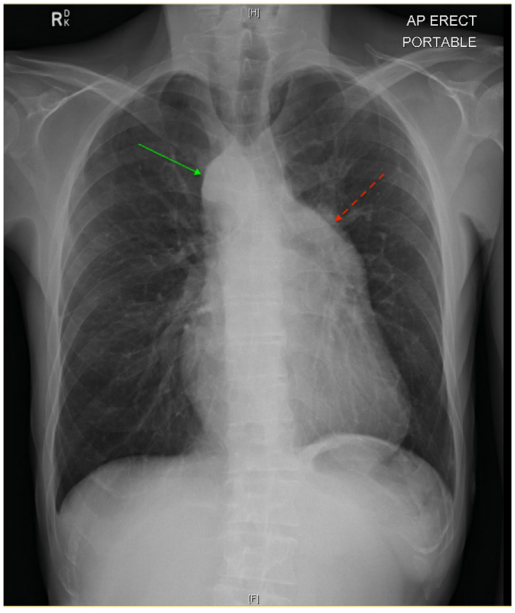
Tricuspid annular calcification is rare, and previously reported cases have almost exclusively been in patients with pulmonary stenosis or large atrial septal defects.[3] In these cases, and ours, severely elevated RV pressure and volume overload from birth likely led to an accelerated process of degeneration and calcification analogous to that more commonly seen

**FIGURE 3 Multi-Modality Cardiac Imaging of Aorta-Pulmonary Window**




Large aorto-pulmonary defect **(arrow)** formed by continuity between main pulmonary artery (MPA) and mid-ascending aorta (Asc Ao), shown on cardiac magnetic resonance imaging **(A)** and computed tomography cardiac chambers **(B)**.

**FIGURE 4 Chest X-Ray**



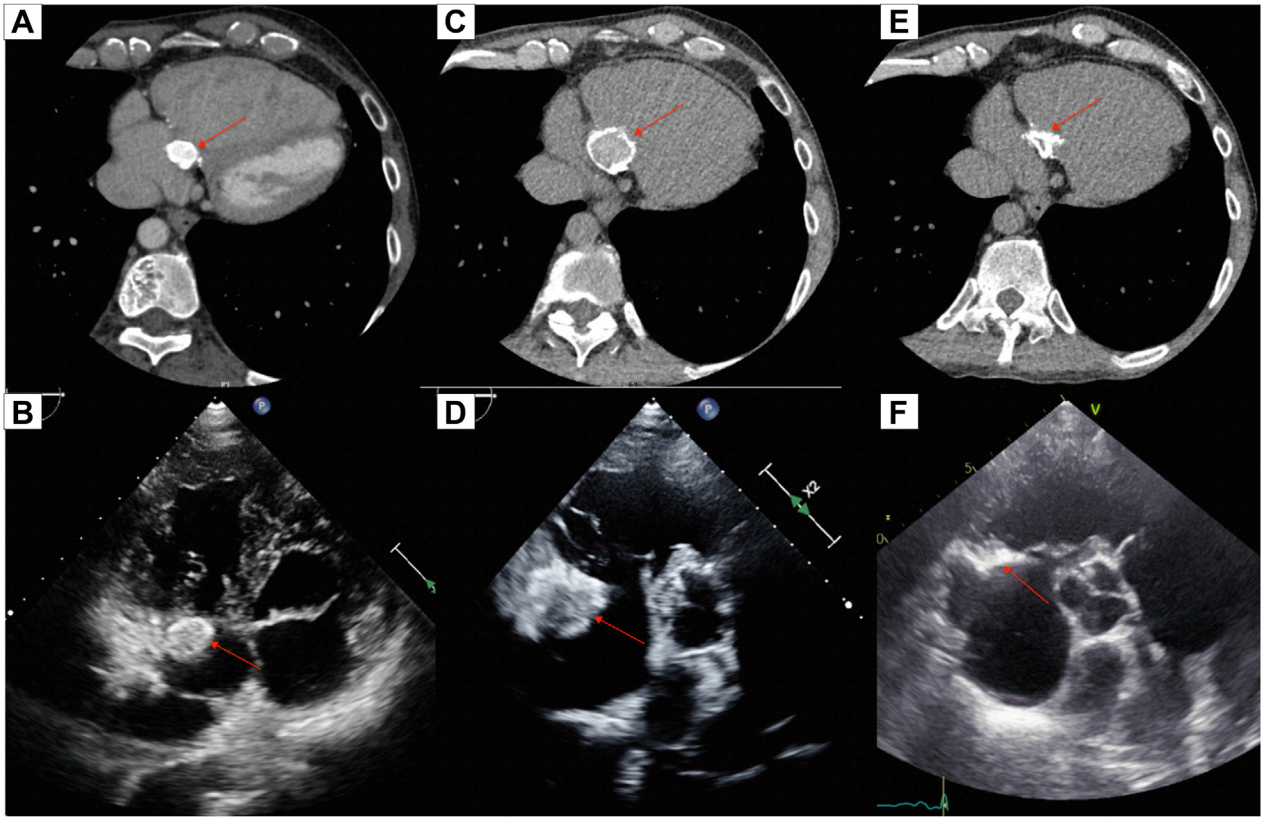

Chest x-ray showing enlarged pulmonary trunk **(dotted arrow)**, central pulmonary plethora, and right-sided aortic arch **(solid arrow)**.

with systemic hypertension and mitral annular calcification.[4]

Multimodality imaging is essential to differentiate caseous tricuspid annular calcification from other cardiac masses, such as vegetations or tumors. The liquefied core of cholesterol and fatty acids produces a central echolucency on echocardiography and corresponding hypodensity on CT imaging. The peripheral calcific rim evident on echocardiography can be confirmed on CT imaging to encircle the valve annulus, with avascularity and lack of enhancement as key distinguishing features.[5]

Pulmonary vasodilator therapies have demonstrated improvement in functional capacity and pulmonary hemodynamic in patients with ES.[6] Our patient initially had a favorable response to Sildenafil with stabilization of symptoms and progressive decline in pulmonary pressures after commencing therapy. Serial imaging demonstrated a temporal correlation between regression of our patient's caseating mass and his worsening pulmonary hypertension with associated clinical decompensation. We hypothesize that this likely reflects exudation of caseating material into the pulmonary arterial circulation, a novel pathophysiologic process that has not been previously documented.

**FIGURE 5** Serial Transthoracic Echocardiograms and Computed Tomography Cardiac Chambers




Serial computed tomography cardiac chambers and transthoracic echocardiograms showing initial finding of tricuspid annular mass **(arrow) (A and B)**, enlargement on repeat imaging **(arrow) (C and D)**, and regression on follow-up **(arrow) (E and F)**.

## CONCLUSIONS

In summary, we report the complex clinical course of one of the oldest surviving adults with ES secondary to uncorrected APW. Our case shows how the markedly deranged physiology in this population can produce unusual complications, such as dynamic tricuspid annular caseation with pulmonary embolization.

## FUNDING SUPPORT AND AUTHOR DISCLOSURES

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

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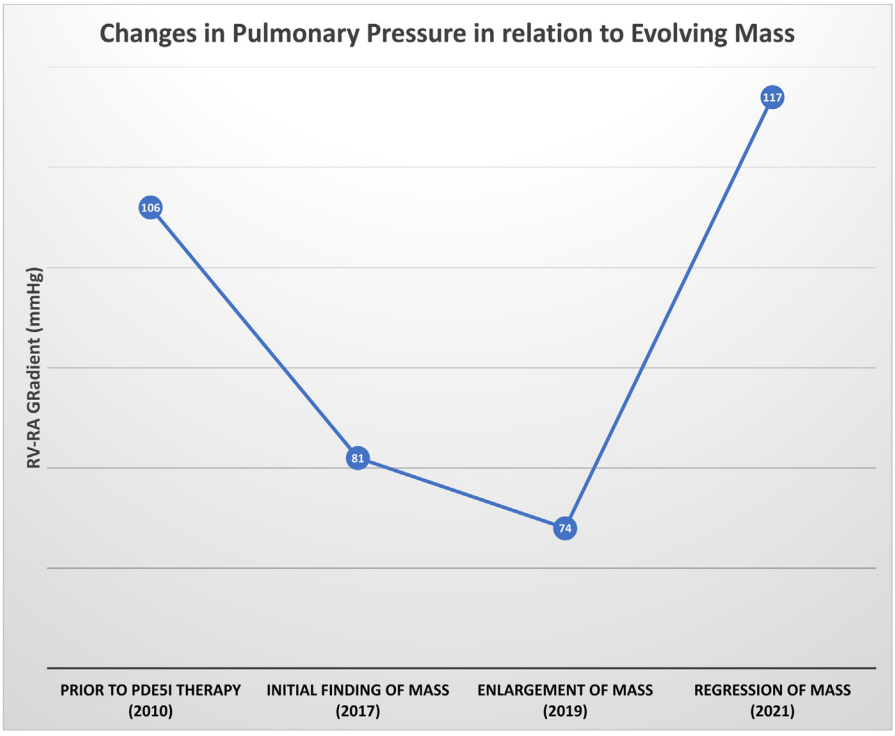
**FIGURE 6** Line Graph of Pulmonary Pressure Over Time




Changes in pulmonary pressures in relation to phosphodiesterase type-5 inhibition (PDE5i) therapy and evolution of tricuspid mass. RV-RA = right ventricle-right atrium.

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**KEY WORDS** aorto-pulmonary window, cerebral abscess, congenital heart disease, Eisenmenger syndrome, pulmonary hypertension, tricuspid annular calcification

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