

# A case report of right ventricular outflow tract obstruction caused by B-cell lymphoma: a rare presentation in an adult patient with pulmonary atresia

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Received 11 June 2021; first decision 12 July 2021; accepted 4 October 2021; online publish-ahead-of-print 7 October 2021

## Background

Right ventricle outflow tract (RVOT) dysfunction is a common long-term complication in adult patients with pulmonary atresia/ventricular septal defect (PA/VSD). Common causes include valve thrombosis, stent fractures, and graft calcification. We present, to the best of our knowledge, the first case of malignant invasion of a Gore-Tex conduit, causing severe right ventricle (RV) failure.

## Case summary

A 30-year-old woman with a history of PA/VSD with major aortopulmonary collateral arteries (MAPCAs) presented with worsening dyspnoea and exercise intolerance. In infancy, she underwent unifocalization of the right- and left-sided AP collaterals utilizing an 18 and 16 mm Gore-Tex graft, respectively. At age 7, she had surgical repair with VSD patch closure and placement of a 20 mm right ventricle-pulmonary artery (RVPA) homograft connected to a 20 mm Gore-Tex graft with linkage to the previously placed right and left unifocalization grafts. A transthoracic echocardiogram revealed a severely dilated RV and a heavily calcified RVOT conduit with severe stenosis. Cardiac computed tomography showed a stenotic RVPA conduit with calcified mural mass. She underwent surgical revision of the RVPA conduit with thromboendarterectomy of bilateral pulmonary arteries. Pathology of the removed conduit revealed fibrin-associated Epstein–Barr virus-positive diffuse large B-cell lymphoma (FA DLBCL).

## Discussion

One prior case report has demonstrated invasion of DLBCL involving an aortic synthetic tube graft. However, malignant invasion of the RVOT Gore-Tex conduit has yet to be reported. Pathological review can be essential in guiding management. Malignant invasion of Gore-Tex conduits is a rare phenomenon, but one that should be closely monitored following repair of the RVOT.

## Keywords

RVOT dysfunction • Gore-Tex conduit • Diffuse large B-cell lymphoma • Pulmonary atresia • Case report

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Handling Editor: Michel Pompeu Sá

Peer-reviewers: Christoph Sinning and Marcelo Haertel Miglioranza

Compliance Editor: Daniel Tardo

Supplementary Material Editor: Ross Thomson

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## Learning points

- Right ventricle (RV) outflow tract obstruction is a common long-term complication in patients with pulmonary atresia/ventricular septal defect and tetralogy of Fallot. It is commonly caused by valve thrombosis, stent fractures, graft calcification, and rarely malignant invasion of the RV to pulmonary artery conduit.
- Various imaging modalities can highlight the extent of graft stenosis, but obtaining intra-operative specimens of the mural mass remains crucial in guiding prompt and appropriate management.

## Introduction

Right ventricle outflow tract (RVOT) dysfunction is a common long-term complication in adult patients with pulmonary atresia/ventricular septal defect (PA/VSD). Common causes include valve thrombosis, stent fractures, and graft calcification. We present, to the best of our knowledge, the first case of malignant invasion of a Gore-Tex conduit, causing severe right ventricle (RV) failure.

## Timeline

### Timeline

<b>Infancy:</b>	Patient was born with a PA/VSD and underwent unifocalization of the right and left sided aortopulmonary (AP) collaterals.
<b>Age 7:</b>	Underwent surgical repair with VSD patch closure and placement of a 20mm RVPA homograft connected to a 20mm Gore-Tex graft with linkage to the previously placed right and left unifocalization grafts.
<b>Day 1:</b>	A 30-year-old female presented with increased lethargy and shortness of breath for three months. She underwent CT cardiac angiogram (congenital heart disease) which revealed an extensive irregular noncalcified mural plaque or thrombus involving the distal main and proximal right and left pulmonary artery conduits, causing severe lumen stenosis. The patient was started on milrinone for RV failure and heparin drip for RVPA homograft stenosis with thrombosis and calcification.
<b>Day 2:</b>	Evaluated by Cardiothoracic surgery who recommended surgical intervention to address the above noted complications. Echocardiogram revealed an ejection fraction of 55-60%, severely dilated right ventricle, and a heavily calcified right ventricle outflow tract (RVOT) conduit with severe stenosis (peak outflow velocity 4.2 m/sec, peak outflow gradient 70-80 mm Hg). RVSP estimated 90-100 mm Hg.
<b>Day 7:</b>	Patient underwent successful surgical intervention with CT surgery including reconstruction of the right ventricle to branch pulmonary artery with a #26 pulmonary homograft, bilateral pulmonary artery thromboembolectomies, and excision of the calcified aortic homograft.
<b>Day 10:</b>	The patient continued to require epinephrine and milrinone drip to augment cardiac function.
<b>Day 13:</b>	Weaned off all pressors.
<b>Day 16:</b>	Discharged home on Warfarin.
<b>Day 21:</b>	Pathology of the removed conduit revealed fibrin associated Epstein-Barr Virus (EBV) positive diffuse large B-cell lymphoma (FA DLBCL) with similar cytological findings in the distal PA thrombus.
<b>&gt;3 months since hospitalization:</b>	Patient underwent successful fluoroscopic guided bone marrow biopsy. Repeat TTE showed favorable RV reverse remodeling with improvement in RV systolic function and normal RVOT hemodynamics with no significant stenosis (peak velocity < 1.5 m/sec).
<b>&gt;5 months since hospitalization:</b>	Chemotherapy R-CVP initiated.

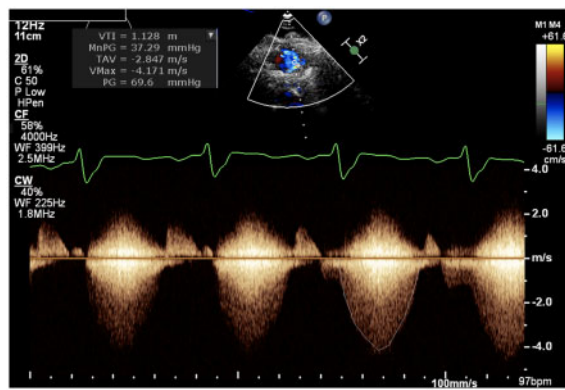
## Case presentation

A 30-year-old woman with congenital heart disease consisting of PA/VSD with major aortopulmonary collateral arteries presented with worsening dyspnoea and progressive exercise intolerance. In infancy, she underwent surgical repair consisting of unifocalization of the right- and left-sided AP collaterals utilizing an 18 and 16 mm Gore-Tex graft, respectively. At age 7, she underwent surgical repair with VSD patch closure and placement of a 20 mm RVPA homograft connected to a 20 mm Gore-Tex graft with linkage to the previously

placed right and left unifocalization grafts. On presentation, she was normotensive and slightly tachycardic (heart rate: 110 b.p.m.). There was a normal first heart sound followed by a high-pitched ejection systolic murmur (4/6) at the left parasternal border. The second heart sound was normal. Elevated neck veins with estimated central venous pressure at 15 cm H<sub>2</sub>O were also noted. Laboratory evaluation revealed slight monocytosis at 12.2% (reference: 2–10%) and elevated prohormone of brain natriuretic peptide at 4843 pg/mL (reference: 30–125 pg/mL).

A variety of imaging modalities were utilized to further investigate the clinical presentation. A transthoracic echocardiogram (TTE) demonstrated a severely dilated RV with severe hypertrophy and moderately reduced systolic function. Left ventricular systolic function was normal with an estimated ejection fraction of 55–60%. The RV systolic pressure was severely elevated near systemic pressures, estimating ~90–100 mmHg based on tricuspid regurgitation Doppler signal. The RVPA conduit was severely calcified with severe RVOT

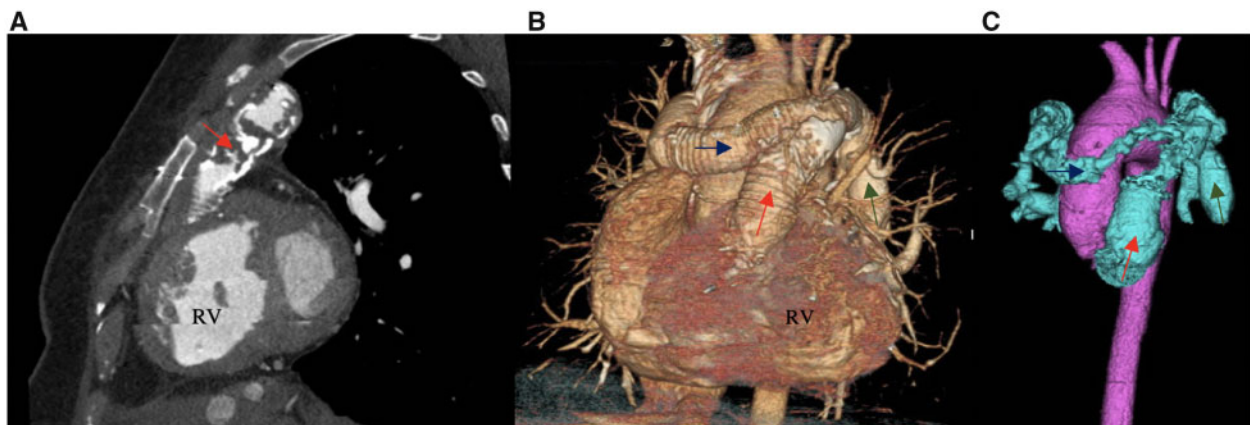
stenosis (peak outflow velocity of 4.2 m/s and peak outflow gradient of 70–80 mmHg) (Figure 1). Cardiac computed tomography (CT) was performed and revealed a stenotic RVPA conduit by a calcified mural mass with subsegmental pulmonary emboli (Figure 2A–C). Given the presentation of RV failure caused by the dysfunctional RVPA conduit, surgical repair was pursued. Intraoperatively, extensive calcification was noted of the RVPA conduit with severe stenosis and erosion into the sternum and left lung. The Gore-Tex tube used to connect the RVPA homograft to the right and left pulmonary artery was full of clots, organized calcium deposits, and atheromas (Figure 3). The patient underwent surgical RVPA conduit revision with thromboendarterectomy of bilateral PAs (Figure 4A and B). Pathology of the removed conduit revealed fibrin-associated Epstein-Barr virus (EBV)-positive diffuse large B-cell lymphoma (FA DLBCL) with similar cytological findings in the distal PA thrombus (Figure 5). Positron emission tomography scan showed increased activity in the lymph nodes and left upper lobe of the lung. With the increased concern



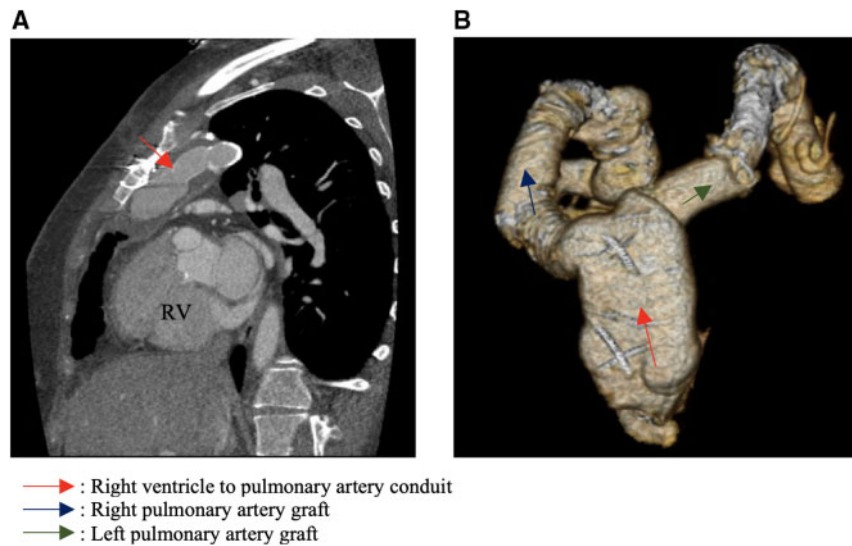
**Figure 1** Peak outflow velocity of 4.2 m/s and peak outflow gradient of ~70 mmHg.



**Figure 3** Fragments of shunt and associated tissue.



**Figure 2** (A) Heavy calcification of the main pulmonary artery conduit with extensive mural hypodense plaque. (B) Three-dimensional view showing heavy calcification of the main, right, and left pulmonary artery conduits with extensive mural hypodense plaque. Dilated right ventricle. (C) Three-dimensional view displaying extensive calcification of the main, right, and left pulmonary artery conduit. RV, right ventricle.

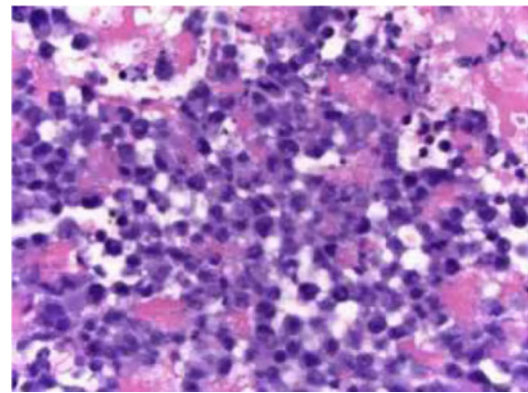


**Figure 4** (A) Sagittal view displaying patent conduit post-RVPA conduit revision with thromboendarterectomy of bilateral pulmonary arteries. (B) Three-dimensional view displaying improvement in conduit calcification. RV, right ventricle.

for haematogenous and lymphomatous emboli in the presence of subsegmental pulmonary emboli, systemic chemotherapy was initiated. Seven rounds of R-CVP (rituximab, cyclophosphamide, vincristine sulfate, and prednisone) chemotherapy were successfully completed with repeat imaging showing resolution of the previously noted increased metabolic activity in the cervical lymph nodes. Follow-up TTE at 3 months revealed favourable RV reverse remodeling with improvement in RV systolic function and normal RVOT haemodynamics without significant stenosis (peak velocity < 1.5 m/s).

## Discussion

This case represents a rare clinical presentation of lymphomatous invasion of an RVPA conduit in an adult patient with PA/VSD. Right ventricle outflow tract dysfunction is a common long-term complication encountered in adult patients with PA/VSD and tetralogy of Fallot. To the best of our knowledge, this is the first case of RVOT dysfunction in an adult patient with PA/VSD caused by malignant invasion of a Gore-Tex conduit causing severe RV failure. RVPA conduit stenosis in adult patients with PA/VSD is commonly caused by calcification, thrombosis, infective endocarditis, or stent fracture, while lymphomatous invasion is rare.<sup>1,2</sup> Invasion of cardiac prosthesis by DLBCL has been described in a few case series,<sup>3</sup> but malignant invasion of the RVOT Gore-Tex conduit, a product of polytetrafluoroethylene, has yet to be reported. Various imaging modalities can highlight the extent of graft stenosis but obtaining intra-operative specimens of the mural mass remains crucial in guiding prompt and accurate management. In our case, cardiac CT demonstrated a filling defect with a preoperative differential diagnosis including extensive calcification and thrombosis. However, malignant invasion was not considered given the rare occurrence. Post-operative pathological examination confirmed FA DLBCL, prompting an interdisciplinary approach in the management.



**Figure 5** Atypical lymphoma cells with irregular nuclear contours, vesicular to marginated chromatin, prominent nucleoli, and scant cytoplasm.

FA DLBCL is composed of large malignant B cells, which are infected by EBV. This particular lymphoma is often found in body cavities or narrow spaces including avascular fibrin masses, blood clots, cysts, or in the vicinity of prosthetic devices.<sup>4</sup> FA DLBCL is typically discovered incidentally during removal of fibrinous material and favours a good prognosis, often requiring only surgical intervention.<sup>4</sup> On occasion, a combination of surgical intervention and systemic chemotherapy can be pursued. Although the pathogenesis of FA DLBCL is not fully understood, the involvement of EBV infection and foreign material are possible contributing factors.<sup>4</sup> In summary, malignant invasion of an RVPA conduit in adult congenital heart disease patients is a rare phenomenon that may cause RVOT dysfunction and RV failure. A high index of suspicion is required for timely recognition and management to ensure favourable outcome.



## Lead author biography



Payush Chatta is an internal medicine resident in training at Loma Linda University Medical Center. He earned his Bachelors in Science from the University of California Los Angeles after which he attended medical school at A.T. Still University School of Osteopathic Medicine in Arizona. There he was a member of Sigma Sigma Phi, a National Osteopathic Honors Society. He ultimately wishes to pursue cardiology

fellowship and further subspecialize as an advanced heart failure cardiologist.

## Supplementary material

[Supplementary material](#) is available at *European Heart Journal - Case Reports* online.

**Slide sets:** A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

**Consent:** The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

**Conflict of interest:** None declared.

**Funding:** None declared.

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