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VASCULAR MEDICINE

CLINICAL CASE

Pulmonary Endarterectomy for Chronic Thromboembolic Pulmonary Hypertension Secondary to Expanded Polytetrafluoroethylene Valved Pulmonary Conduit Thrombosis

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ABSTRACT

BACKGROUND There is no consensus on the preferred conduit for right ventricular outflow tract (RVOT) reconstruction for congenital cardiac disease.

CASE SUMMARY We present a case of a 21-year-old woman with history of tetralogy of Fallot presenting with recurrent graft thrombosis and pulmonary emboli in the setting of RVOT reconstruction with a 20-mm expanded polytetrafluoroethylene (ePTFE) valved conduit (GORE PV1, W.L. Gore & Associates). A diagnosis of chronic thromboembolic pulmonary hypertension was made, and the patient underwent pulmonary endarterectomy and conduit exchange with a pulmonary homograft.

DISCUSSION The novelty and clinical significance of this report lies in the confirmation of thrombosis of a large-diameter ePTFE valved pulmonary conduit (20 mm) and its implications for monitoring and need for further optimization of thromboprophylaxis strategies in this population. (JACC Case Rep. 2025;30:103069) © 2025 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

Our case was a 21-year-old woman born with tetralogy of Fallot and an absent pulmonary valve with left pulmonary artery (PA) takeoff from the ductus arteriosus. At 7 days of age, she underwent insertion of a

TAKE-HOME MESSAGE

- Thrombosis of an ePTFE valved pulmonary conduit can lead to CTEPH and warrants conduit exchange at the time of pulmonary thromboendarterectomy.

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ABBREVIATIONS AND ACRONYMS

CTEPH = chronic thromboembolic pulmonary hypertension

CTPA = computed tomography of the pulmonary artery

ePTFE = expanded polytetrafluoroethylene

PA = pulmonary artery

RVOT = right ventricular outflow tract

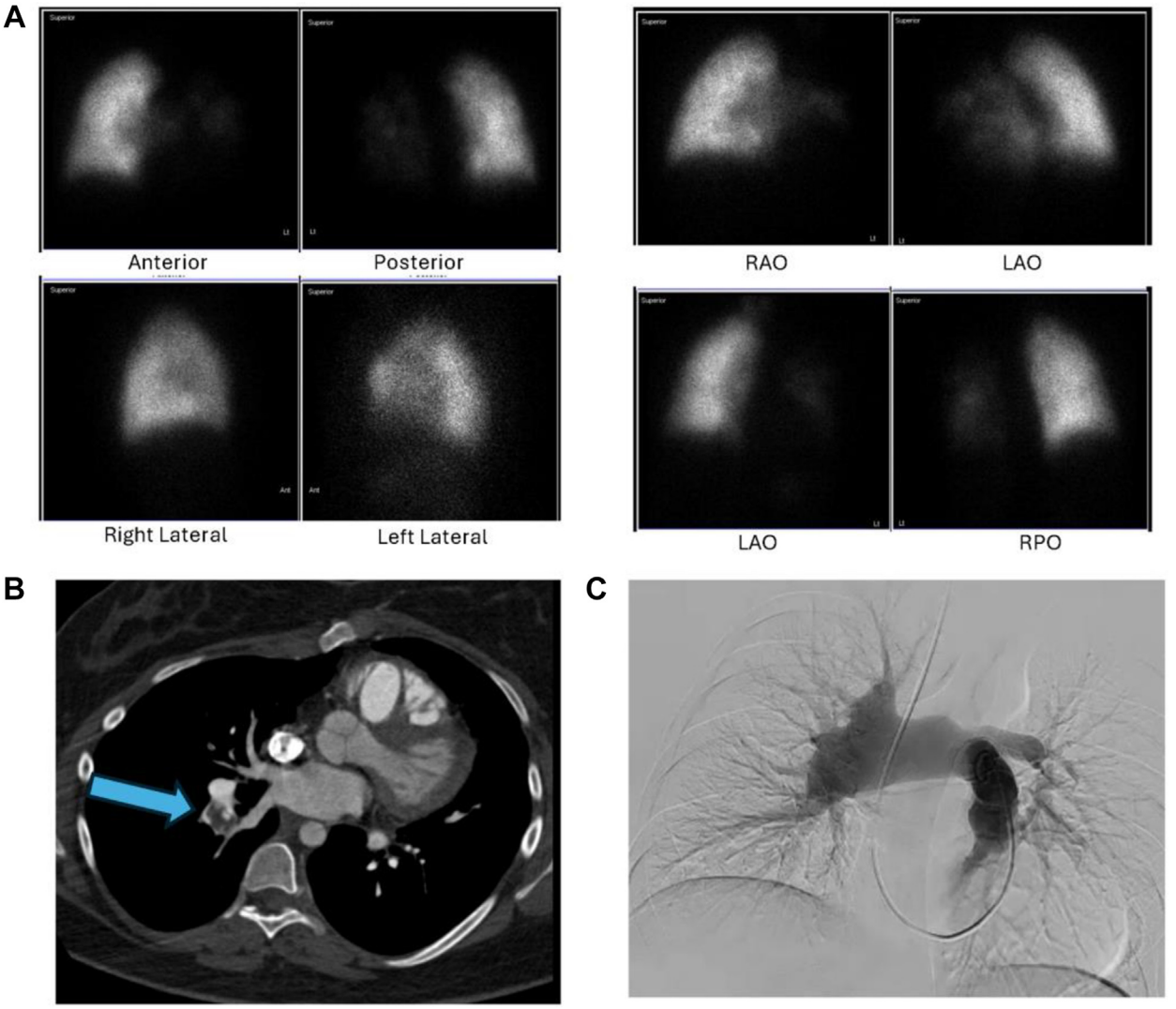
WU = Wood units

right ventricle to PA 11-mm pulmonary homograft, ventricular septal defect closure, right PA plication, and arterioplasty of the left PA with reimplantation. At 7 months of age, she underwent exchange of the pulmonary conduit with a 15-mm pulmonary homograft and revision of the left PA anastomosis. At age 16 years, she underwent a third-time pulmonary conduit exchange with a 20-mm expanded polytetrafluoroethylene (ePTFE) valved conduit (GORE PV1; W.L Gore & Associates), right PA plication, and left PA arterioplasty. The procedure was uncomplicated with only trivial regurgitation noted on a transesophageal echocardiography. She was extubated postprocedure and discharged on postoperative day 5 on a 6-month course of 325 mg of aspirin.

PAST MEDICAL HISTORY

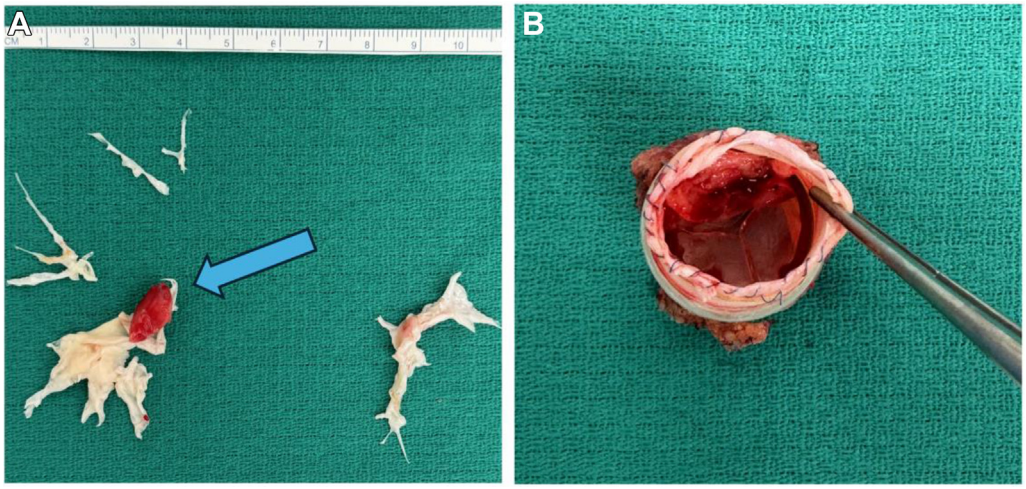
The patient's past medical history includes Chiari malformation type 1 and scoliosis.

FIGURE 1 Diagnostic Evaluation for Chronic Thromboembolic Pulmonary Hypertension



(A) Quantitative lung perfusion scintigraphy demonstrating asymmetrical perfusion with 11% to the left lung and 89% to the right lung. (B) Computed tomography pulmonary angiogram demonstrating bilateral thromboembolic disease (arrow) and dilation of the proximal right pulmonary artery (20.0 × 22.8 mm). (C) Pulmonary artery catheterization and angiogram with mean pulmonary artery pressure (21 mm Hg) and pulmonary vascular resistance (2.3 WU).

FIGURE 2 **Pulmonary Endarterectomy and ePTFE Conduit Specimen**



(A) Endarterectomy specimen with thickened intima and fibrous webs and bands as well as evidence of more acute thromboembolic disease (arrow). (B) En-bloc ePTFE conduit with pathologically confirmed organized thrombus at the site of distal anastomosis. ePTFE = expanded polytetrafluoroethylene.

INVESTIGATIONS

Routine cardiac magnetic resonance imaging performed approximately 2.5 years postprocedure demonstrated an unbalanced differential pulmonary blood flow of 90% to the right lung and 10% to the left lung by 2- and 4-dimensional flow (compared with 60% to the right lung and 40% to the left lung preprocedure). She underwent elective right heart catheterization and was noted to have thrombus in the left PA. Suction thrombectomy was attempted, and balloon angioplasty was performed. A postprocedure computed tomography of the PA (CTPA) demonstrated an asymmetrical thin crescentic low-attenuation material in the mid conduit consistent with thrombus as well as a nonocclusive thrombus in the distal left PA, right interolobar PA, and in bilateral segmental arteries (**Figure 1**).

DIFFERENTIAL DIAGNOSIS

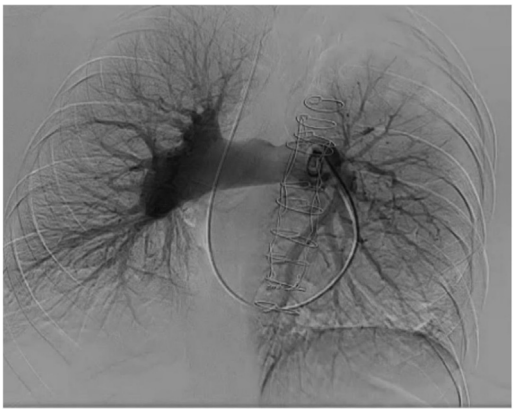
Common causes of valved conduit failure include endocarditis, degeneration and calcification, technical issues during implantation, and pulmonary insufficiency.[1]

MANAGEMENT

She was started on rivaroxaban 20 mg. A repeat CTPA at 5 months demonstrated progression with a new thrombus at the distal anastomosis of the valved conduit. She was then transitioned to warfarin with an international normalized ratio goal of 2.0 to 3.0. This was increased to an international normalized ratio goal of 2.5 to 3.5 after a follow-up CTPA demonstrated concern for progression. There was concern the conduit was thrombogenic, and the patient was referred to the chronic thromboembolic pulmonary hypertension (CTEPH) program with a multidisciplinary recommendation for pulmonary endarterectomy and conduit exchange. She was exhibiting NYHA functional class III symptoms with a mean PA pressure of 21 mm Hg, pulmonary vascular resistance 2.3 WU, cardiac output of 3.4 L/min, and a cardiac index of 2.02 L/min.

The patient underwent a median sternotomy for a fourth time with the conduit excised en bloc (Video 1). Bilateral pulmonary endarterectomy was performed with the patient under deep hypothermic circulatory arrest (18 °C) with 2 circulatory arrests and augmentation of the left PA. Right ventricular trabeculations

FIGURE 3 Pulmonary Artery Catheterization and Angiogram 3 Months Postprocedure



The study demonstrated improved pulmonary vascular resistance (0.89 WU).

were excised to augment the right ventricular outflow tract (RVOT), and a 28-mm pulmonary homograft (LifeNet Health Cardiograft) was implanted (Figure 2). Bypass time was 250 minutes, and aortic cross-clamp time was 105 minutes. Total hypothermic circulatory arrest time was 31 minutes (18 minutes right, 13 minutes left). She was extubated on postoperative day 1, transferred from the cardiothoracic intensive care unit on postoperative day 2, and discharged on postoperative day 5 on warfarin with an international normalized ratio goal of 2.5 to 3.5.

DISCUSSION

There is no consensus on the preferred conduit for RVOT reconstruction. A range of reconstructive options exist including a homograft (pulmonary or aortic) and xenograft as with the Contegra, a valved bovine jugular conduit from Medtronic. Bioprosthetic options include the Hancock Dacron tube graft containing a porcine valve (also from Medtronic) and ePTFE Gore-Tex conduits (W.L. Gore & Associates). The various conduits have been shown to differ across short- and long-term outcomes in rate of failure and reintervention.[2] The long-term shelf stability and availability are attractive features of synthetic conduits.

In a review of the literature, we identified 1 report of thrombus associated with ePTFE reconstruction of the RVOT in a 46-year-old woman who underwent transannular patch angioplasty at age 8 years.[3] The ePTFE-based valved conduit (GORE PV1 device) discussed in this case was subject to a prospective, multicenter, single-arm, early feasibility study to evaluate the safety and performance of the device.[4] In the published preliminary results, thrombus was suspected in 1 patient (9.1%) based on cardiac magnetic resonance imaging but was not appreciated on echocardiography. Anticoagulation therapy was not protocolized in this study. Several studies have demonstrated comparable antithrombotic effects between antiplatelet agents and anticoagulation therapy. Aspirin monitoring and the use of direct oral anticoagulants represent potential future directions in this population.[5]

Therefore, the novelty and clinical significance of this report lies in the confirmation of thrombosis of a large-diameter ePTFE valved pulmonary conduit (20 mm) and its implications for monitoring and anticoagulation strategies in this population.[5,6] Kim et al reported 1 instance of acute thrombus associated with an ePTFE biscuspid pulmonary valve with extension to the left PA.[7] The authors noted the presence of acute angulation of the left PA in that case. It is also important to note that tetralogy of Fallot with absent pulmonary valve and nonconfluent PA is an uncommon constellation with only a handful of case reports on its surgical management and limited data on lifetime management.[8] Congenital PA anomalies may result in more turbulent conduit flow and increase the risk of conduit thrombosis.[9] Finally, our report also demonstrates the safety and tolerability of pulmonary endarterectomy performed with concomitant RVOT reconstruction and highlights the value of long-term surveillance and management of patients with congenital cardiac disease at a center with an affiliated CTEPH program.

FOLLOW-UP

At 3 months postdischarge, pulmonary vascular resistance was 0.89 WU (from a preprocedure 2.3 WU), mean PA pressure of 20 mm Hg, cardiac output of 4.47 L/min, and cardiac index of 2.64 L/min. Residual CTEPH was appreciated in left-sided segments 9A and 10A (Jamieson III and IV) (Figure 3). The patient's 6-minute walking distance was 360 m (Borg 0-2; no supplemental oxygen; O_2 100% to 97%) compared with 420 m (Borg 0-3; no supplemental oxygen; O_2 99% to 98%). At the 5-month follow up, her symptoms had improved to NYHA functional class II from NYHA class III preprocedure, with residual exertional chest pain. A CTPA at 9 months was

without evidence of new pulmonary emboli or conduit thrombosis.

CONCLUSIONS

Thrombosis is an acute and long-term complication of an ePTFE valved pulmonary conduit with potential for progression to CTEPH. This complication warrants a high index of suspicion and further study of potential contributing factors and optimization of anticoagulation strategies in this population.

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KEY WORDS chronic thromboembolic pulmonary hypertension, pulmonary conduit, tetralogy of Fallot

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