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CASE REPORT: CLINICAL CASE: ACC.24

Total Thrombotic Occlusion of an Extracardiac Fontan Conduit in a Hemodynamically Stable Patient

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ABSTRACT

Cardiac and extracardiac complications of single-ventricle physiology are well described and carry significant morbidity and mortality risks. We present the case of a 32-year-old patient with a Fontan circulation who presented to the emergency department with epigastric pain, and with chest computed tomography angiography concerning for total thrombosis of the extracardiac conduit. JACC Case Rep 2024;29:102421) © 2024 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

A 32-year-old woman born with complex single-ventricle anatomy, including a complete, right-dominant atrioventricular septal defect with malposed great arteries, pulmonary valve atresia with nonconfluent pulmonary arteries, a hypoplastic left ventricle, an absent right superior vena cava with a left superior vena cava draining to the coronary sinus, and a total anomalous pulmonary venous connection, initially underwent palliation with a Waterson shunt, followed by placement of a right modified Blalock-Thomas-Taussig shunt. She subsequently underwent left bidirectional cavopulmonary anastomosis with repair of the pulmonary veins and ultimately underwent an extracardiac Fontan procedure. The patient presented to the emergency department with epigastric pain radiating to her midchest, midabdomen, and back that was associated with dyspnea on exertion, nausea, and lightheadedness. The patient was otherwise hemodynamically stable, with normal vital signs at presentation and with her baseline oxygen saturation in the mid-90s. There were no signs of respiratory distress, and she had normal

LEARNING OBJECTIVES

- To have a high index of suspicion for thromboembolic events despite having low clinical suspicion.
- To assess for thromboembolic complications adequately with the aid of multimodality imaging.
- To assess for additional cardiac or extracardiac risk factors that will increase the potential of a thrombotic complication.

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ABBREVIATIONS AND ACRONYMS

DOAC = direct oral anticoagulant

DVT = deep vein thrombosis

mentation. The physical examination was otherwise at baseline, with a mild degree of abdominal distention.

PAST MEDICAL HISTORY

Given the patient s nonocclusive thrombosis and stenosis in the Fontan conduit that was unresponsive to transcatheter stent placement, she underwent an 18-mm extracardiac conduit replacement at 13 years of age that had a slight residual bend in the midportion with no evidence of obstruction by previous diagnostic studies. Furthermore, 2 months before her current presentation she had left lower extremity edema and was found to have deep vein thrombosis DVT), for which she was started on a direct oral anticoagulant DOAC); however, as a result of noncompliance, she stopped taking the medication after 1 month.

DIFFERENTIAL DIAGNOSIS

Conditions to consider, given her clinical description, included gastrointestinal causes gastritis, gastroesophageal re ux disease, or pancreatitis), a musculoskeletal source, and even though less likely from a cardiovascular perspective, aortic dissection or acute coronary syndrome. Given the presence of a Fontan circulation and a history of DVT, pulmonary embolism and thromboembolic events were also considered as a possible cause for her epigastric and chest pain despite having low clinical suspicion.

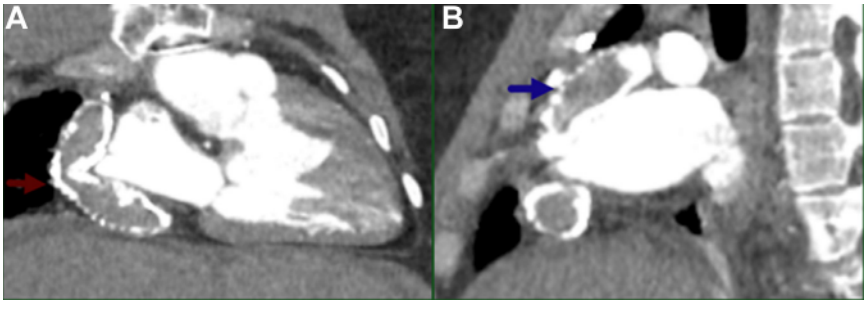
INVESTIGATIONS

Laboratory studies were remarkable for an elevated D-dimer of 5,840 ng/mL. Additional laboratory studies included a complete blood count, a comprehensive metabolic panel, and troponin levels, all of which were within normal range. Imaging included bilateral ultrasound scans of the lower extremities, which showed no evidence of DVT. An electrocardiogram remained unchanged in comparison with previous studies, with normal sinus rhythm and nonspecific ST-/T-wave changes. A transthoracic echocardiogram demonstrated a dilated right ventricle with mildly depressed systolic function, mild aortic insufficiency, mild atrioventricular valve regurgitation, an unobstructed superior cavopulmonary anastomosis, and a linear echodensity in the inferior vena cava with low- ow velocity. However, the Fontan pathway was not able to be assessed. Ultimately, the patient underwent standard pulmonary embolism protocol chest computed tomography angiography; findings were concerning for filling defects in the distal right pulmonary artery and total thrombosis of the extracardiac conduit that extended to the inferior vena cava with acute angulation at its midportion **Figures 1A 1B and 2A to 2C**).

MANAGEMENT

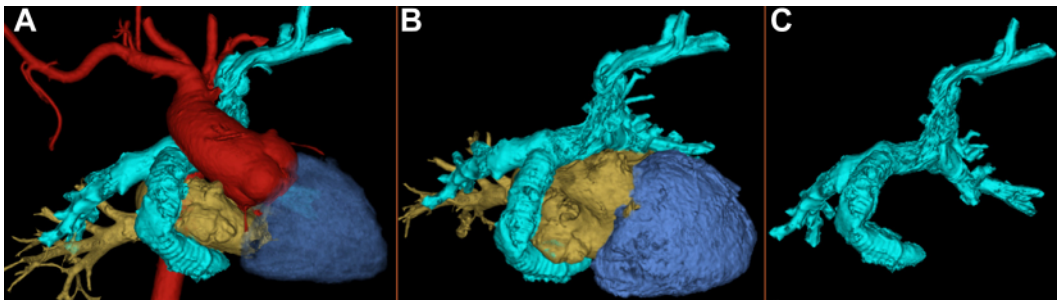
Once admitted, the patient developed significant ascites and recurrent gastrointestinal bleeding. She underwent esophagogastroduodenoscopy, which found moderate portal hypertensive gastropathy, with gastric antral vascular ectasia that was treated with radiofrequency ablation, and grade II esophageal varices, which were banded. Given concerns for thrombosis within the Fontan pathway, the patient underwent cardiac catheterization **Figure 3**), which demonstrated a mean inferior vena cava pressure of 32 mm Hg. She then underwent catheter

FIGURE Computed Tomography Angiography



Computed tomography angiography with delayed phase imaging. A) Coronal view shows nonenhancement within the extracardiac Fontan conduit with an abrupt angulation at its midportion red arrow). B) Sagittal view shows nonenhancement within the extracardiac Fontan conduit with adequate opaci cation of the pulmonary vasculature blue arrow).

FIGURE 2 3 Dimensional Computed Tomography



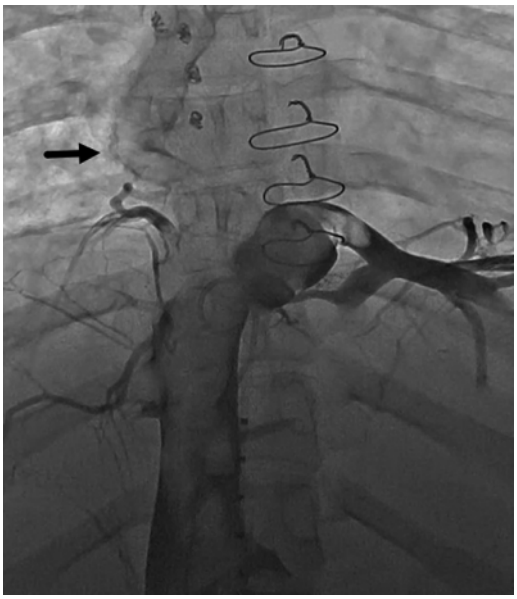
A to C) Three-dimensional computed tomography reconstruction of the extracardiac Fontan conduit that connects to the right pulmonary artery with abrupt angulation at its midportion, as well as a left cavopulmonary anastomosis of the left superior vena cava to the left pulmonary artery light blue). A and B) The spatial relationship with other cardiovascular structures.

thrombectomy with a thromboaspiration system of a large obstructive thrombus in her Fontan pathway and inferior vena cava, followed by stent angioplasty with 2 16 mm by 4.5 cm mounted covered Cheatham-Platinum stents Numed), after which there was improved ow through the conduit. However, despite further stent dilation with a high-pressure balloon at 18 atm, significant angulation at the area of initial obstruction remained, in addition to simultaneous and ongoing thrombus formation even in the presence of therapeutic heparinization. Hence, although the patient was high risk, she ultimately underwent a surgical Fontan conversion from an extracardiac conduit to a lateral tunnel Fontan with an inferior vena cava thrombectomy.

DISCUSSION

Patients with single-ventricle physiology typically undergo staged palliation culminating in a Fontan procedure.[1] Cardiac and extracardiac complications of single-ventricle physiology are well described and include arrhythmias, failure of the Fontan circulation, liver dysfunction, protein-losing enteropathy, conduit stenosis, thrombotic complications, chronic kidney disease, and/or psychological, psychiatric, and cognitive defects, among others.[2] Hemodynamic abnormalities are frequently associated with thrombotic events within the Fontan pathway and carry significant morbidity and mortality risk, often requiring medical or surgical interventions. In addition, early recognition of and intervention for conduit stenosis, a known late complication of the Fontan procedure related to progressive calcification or shape distortion, are paramount to avoid further associated complications.[3] As for the thromboembolic phenomenon, it is partly attributed to an underlying coagulation factor abnormality in addition to unconventional passive and relatively stagnant ow from the systemic venous system to the pulmonary arteries that bypasses a subpulmonary pump.[4] Patients with Fontan palliation who also have atrial tachyarrhythmia are at even higher risk for thrombotic

FIGURE 3 Cardiac Catheterization



Cardiac catheterization with selective injection into the inferior vena cava demonstrating total occlusion of the extracardiac Fontan conduit, as well as abrupt angulation at its midportion black arrow).

complications.[5] It is also important to highlight the need for adequate multi-imaging protocols because the unique anatomical and physiological changes of a Fontan circuit may contribute to differential streaming from the systemic venous system into the branch pulmonary arteries that could result in a false positive diagnosis of a thromboembolic event.[6] Finally, once a significant thromboembolic complication is identified, it is imperative to treat it promptly because this could lead to life-threatening conditions, particularly in the case of substantial conduit thrombosis or pulmonary embolism that acutely diminishes the cardiac preload, thereby compromising the cardiac output.[7]

FOLLOW UP

The patient s postoperative course was complicated by hemodynamically significant atrial and ventricular arrhythmias, sepsis, spontaneous bacterial peritonitis, and cecal perforation requiring emergency ileocecectomy, as well as acute kidney injury with the need for continuous renal replacement therapy that was eventually transitioned to intermittent hemodialysis. Serial postoperative echocardiograms demonstrated an unobstructed Fontan pathway with subjectively low normal right ventricular systolic function. She ultimately was started again on a DOAC and continued with clinical improvement to work on her overall rehabilitation.

CONCLUSIONS

This case demonstrates the need for a high index of suspicion for thromboembolic events in patients with a Fontan circulation, even in the absence of hemodynamic instability or respiratory distress, given the significant associated morbidity and mortality.

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

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