Native Aortic Root Thrombus in 3-Year-Old Fontan Patient with Hypoplastic Left Heart Syndrome: Presentation and Echocardiographic Findings of This Life-Threatening Complication

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INTRODUCTION

Late development of native aortic thrombus after Fontan in children with single ventricle physiology and aortic atresia is a rare and life-threatening complication.[1] In this case report, we describe a 3-year-old patient who acutely presented with third-degree heart block early after completion of Fontan palliation and was subsequently found to have a new aortic root thrombus identified by transthoracic echocardiogram. Transthoracic and transesophageal echocardiography were used to assess for thrombus resolution after therapy and identified redevelopment of the thrombus with clinical deterioration. Echo imaging was invaluable in the diagnosis and management of this patient.

CASE PRESENTATION

A 3-year-old patient with hypoplastic left heart syndrome (HLHS) and mitral stenosis/aortic atresia presented 1 month post–Fontan palliation with new-onset lethargy and vomiting. The patient had previously undergone Norwood palliation with Blalock-Taussig-Thomas shunt as a neonate, with aortic arch reconstruction and creation of the neoaorta (former pulmonary artery) from the single right ventricle to provide systemic output. Pulmonary blood flow was provided through the Blalock-Taussig-Thomas shunt from the subclavian artery to the pulmonary arteries. A bidirectional Glenn was performed at 3 months of age, with takedown of the Blalock-Taussig-Thomas shunt and anastomosis of the superior vena cava to the pulmonary artery, with resultant passive pulmonary blood flow through the superior vena cava. The patient then underwent a nonfenestrated extracardiac Fontan, with anastomosis of the inferior vena cava to the pulmonary artery. The most recent transthoracic echocardiogram obtained at the time of discharge after the patient's Fontan operation demonstrated good right ventricular function, trivial tricuspid valve regurgitation, and a diminutive left ventricular cavity with no thrombus (Figure 1). Parasternal long-axis imaging demonstrated a dilated and apex-forming systemic right ventricle anteriorly, a hypoplastic left ventricle, and a diminutive native ascending aorta.

The patient was noted to be bradycardic, with heart rate in the 30s at a local emergency department, and an electrocardiogram revealed new complete heart block, prompting transfer to our institution. A transthoracic echocardiogram obtained on admission found extensive new thrombus in the diminutive left ventricle, most clearly demonstrated in parasternal long-axis imaging (Figure 2A and Video 1). Parasternal short-axis imaging further delineated new thrombus in the native aortic root near the ostium of the left coronary artery (Figure 2B and Video 2), which was seen extending into the root of the left main coronary artery by two-dimensional echocardiography (Video 3), and color Doppler demonstrated limited anterograde flow (Video 4). The proximal coronary arteries were well visualized with two-dimensional echocardiography using 6 MHz harmonic imaging. Subcostal imaging, with the image inverted and the liver positioned caudally, depicted the thrombus extending into an aortic root diverticulum that was previously seen on pre-Fontan cardiac catheterization (Video 5). Right ventricular systemic function was mild to moderately diminished, which was decreased compared with the patient's post-Fontan discharge echocardiogram 1 month earlier. There was mild tricuspid valve regurgitation, which was consistent with prior echocardiographic imaging. The patient had been managed after the Fontan operation with aspirin and warfarin, and at the time of admission the patient's international normalized ratio was 2.36. Serum troponin was elevated at 34 ng/mL.

The patient care team felt the aortic root thrombosis identified on echocardiogram resulted in acute coronary syndrome secondary to thrombotic coronary occlusion, with atrioventricular nodal ischemia causing heart block and elevated troponin. The patient was then started on systemic thrombolytic therapy with alteplase (tPA) at 0.2 mg/kg/hour. Repeat transthoracic echocardiogram to assess interval changes the following day showed resolution of the aortic root thrombus, but clinical deterioration persisted with abnormal troponin levels, low cardiac output, and elevated Fontan pressures at 20-25 mm Hg. With ongoing complete heart block, the patient was taken to the operating room for epicardial pacemaker placement. In the operating room the patient acutely decompensated and was cannulated onto venoarterial extracorporeal membrane oxygenation (ECMO). Transesophageal echocardiogram (Video 6) showed no

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Conflicts of Interest: None.

Informed consent was obtained from legal guardians, and the legal guardians consented to the case report.

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VIDEO HIGHLIGHTS

Video 1: Transthoracic echocardiogram in the parasternal long-axis view shows organized thrombus filling the hypoplastic left ventricle, with suggestion of thrombus in ascending aorta.

Video 2: Transthoracic echocardiogram in the parasternal short-axis view depicts a well-defined thrombus in the native aortic root near the ostium of the left main coronary artery (*large arrow*).

Video 3: Transthoracic echocardiogram in the parasternal short-axis view demonstrates thrombus in the native aortic root with extension into the left main coronary artery.

Video 4: Transthoracic echocardiogram in the parasternal short-axis view, with the use of color Doppler, demonstrates limited laminar, nonturbulent anterograde flow, seen as *red* in this image moving toward the transducer into the left main coronary artery.

Video 5: Transthoracic echocardiogram in the subcostal coronal/long-axis view with the liver positioned caudally demonstrates thrombus in the native aortic root extending into the native ascending aorta.

Video 6: Transesophageal echocardiogram demonstrates low-velocity filling of the left main coronary artery and distal branches. This could only be demonstrated at a color Doppler Nyquist limit of 0.15 m/sec.

Video 7: Intraoperative angiogram of the native root shows a dilated proximal left coronary artery with severe tapering of the distal branches (*small arrows*) and no anterograde flow to the proximal right coronary artery (*large arrow*). Filling of a diverticulum at the base of the native aortic root is also seen.

Video 8: Transthoracic echocardiogram in the parasternal short-axis view depicts resolution of thrombus in the native aorta and no evidence of thrombus in the proximal right and left main coronary arteries.

Video 9: Transthoracic echocardiogram in the parasternal short-axis view shows easily identified coronary flow in the left main coronary and left anterior descending coronary arteries, which are significantly improved compared with before directed tPA therapy.

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aortic root thrombus and limited blood flow in the left coronary artery. Intraoperative aortic root angiography showed proximal left coronary artery dilation with distal branch tapering, and no flow was seen in the right coronary artery (Figure 3 and Video 7). Because of concerns about persistent coronary artery thrombus as the etiology of the abnormal coronary imaging, a pigtail catheter was placed in the aortic root for catheter-directed tPA administration.

After 3 days of focal thrombolytic therapy, there was spontaneous resolution of complete heart block with conversion to sinus rhythm and improved right ventricular systolic function and increased cardiac output on ECMO. Repeat transthoracic echocardiography (Figure 4, Videos 8 and 9) showed patent coronary arteries with significantly

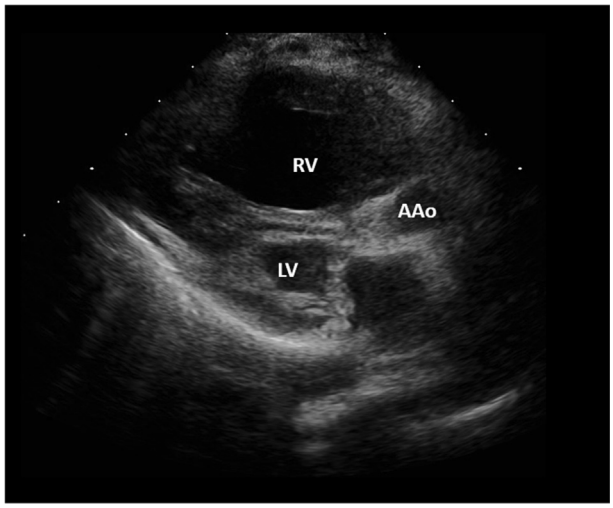

Figure 1 Transthoracic echocardiogram in the parasternal long-axis view obtained on discharge after Fontan procedure demonstrates a diminutive left ventricle (LV) consistent with HLHS, a large right ventricle (RV), and the native ascending aorta (AAo). There is absence of thrombus in the left ventricle and in the native aortic root.

improved anterograde flow into both the left and right coronary arteries. The patient was decannulated from ECMO after 6 days of ECMO support, and direct thrombolytic therapy was again stopped. The aortic root catheter providing access for direct thrombolytic therapy was similarly removed. Systemic anticoagulation was restarted, but in the absence of directed tPA there was recurrence of aortic root thrombus and the patient was placed on mechanical support. The patient was supported with mechanical circulatory support in the form of a single ventricular assist device. Unfortunately, the patient suffered a catastrophic thrombotic neurologic event and subsequently died after 3 weeks of support.

DISCUSSION

Thrombosis is a known complication with single ventricle physiology, predominantly during the first stage of surgical palliation with acute aortopulmonary shunt occlusion.[2,3] In addition, thrombosis and thromboembolic events are known complications in patients with Fontan circulation and are associated with significant morbidity and mortality.[3]

Formation of a native aortic thrombus is described in a few case series[1,4-7] but is a rare and serious complication in patients with HLHS after Norwood palliation, specifically with aortic valve atresia. A review of published case studies reveals that presenting complications associated with native aortic root thrombosis can include thromboembolism, single ventricle dysfunction/heart failure, and conduction abnormalities. Echocardiographic evaluation of the native aortic root is important when patients present with these complications. Visualization of the native aortic root and diminutive left ventricle is usually available in infants and young children with standard parasternal two-dimensional imaging and provides valuable information regarding possible clot formation.

Management strategies include systemic anticoagulation, systemic thrombolysis, catheter-based coronary interventions and discussion with adult interventional cardiology experts, directed thrombolysis, and surgical thromboembolectomy.[8] The use of catheter-directed

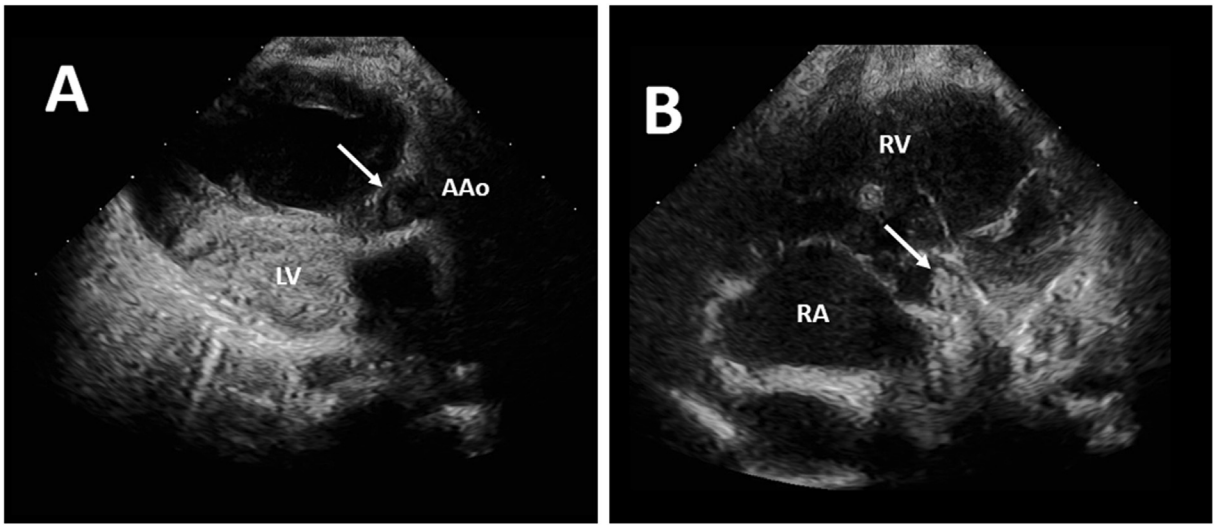

Figure 2 **(A)** Transthoracic echocardiogram in the parasternal long-axis view shows organized thrombus filling the hypoplastic left ventricle (LV) and separate thrombus in the native aortic root (*large arrow*). **(B)** Transthoracic echocardiogram in the parasternal short-axis view shows thrombus (*large arrow*) in the native aortic root near the ostium of the left main coronary artery. *AAo*, Ascending aorta; *RA*, right atrium; *RV*, right ventricle.

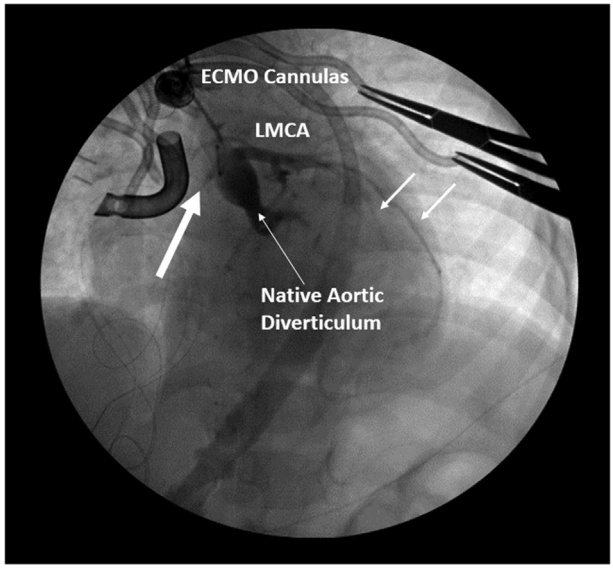

Figure 3 Intraoperative angiography shows a dilated proximal left coronary artery with severe tapering of the distal branches (*small arrows*) and a lack of anterograde flow to the proximal right coronary artery (*large arrow*). A diverticulum at the base of aortic valve is also appreciated. *LMCA*, Left main coronary artery.

tPA, specifically continuous administration outside of the operating room or cardiac catheterization lab, has not been well described in the literature.[9] The hemorrhagic risk associated with tPA is significant in this scenario; however, in the absence of a standard practice to guide management of this critical presentation, the use of tPA was felt to be worth the therapeutic benefit in this patient's case. Although our patient ultimately died due to complications of a single ventricle assist device with low cardiac output and multiorgan system failure, directed tPA therapy was effective in reducing the thrombotic burden and should be considered early when a patient presents with signs of coronary compromise. Transthoracic and transesophageal echocardiography can be effective imaging modalities to assess for interval changes in response to therapy.

CONCLUSION

Echocardiographic imaging of the native aortic root patency is important in patients with HLHS with aortic atresia after surgical palliations. New-onset complications suggestive of thromboembolism and/or coronary insufficiency should result in focused native aortic root

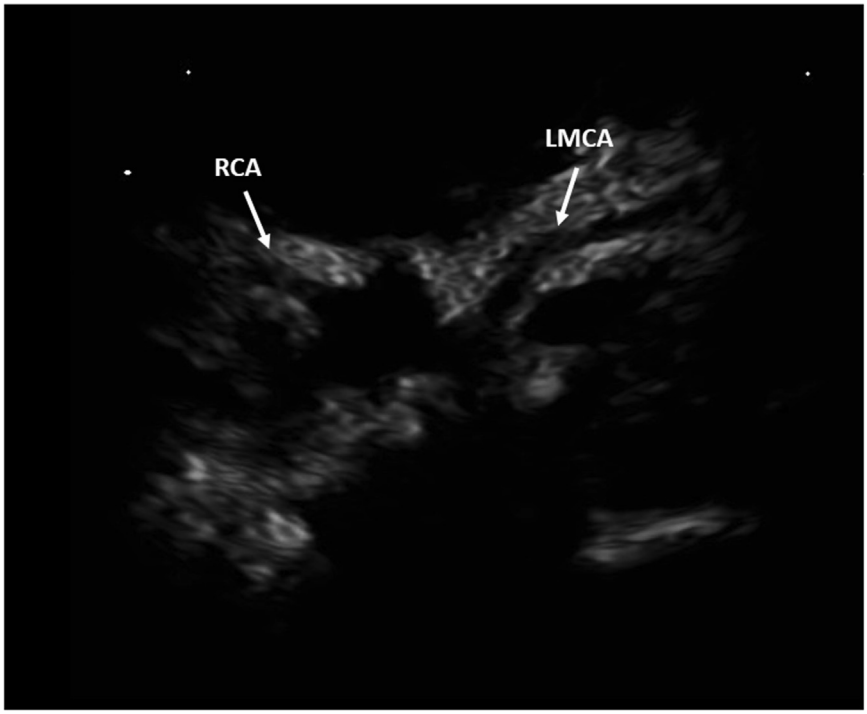

Figure 4 Transthoracic echocardiogram in the parasternal short-axis view demonstrates patent left and right coronary arteries after the use of catheter-directed tPA. *LMCA*, Left main coronary artery; *RCA*, right coronary artery.

imaging to rule out thrombosis as a cause in this subset of patients, and echocardiography can be a useful tool to help guide thrombolytic management strategies.

SUPPLEMENTARY DATA

Supplementary data to this article can be found at https://doi.org/10.1016/j.case.2021.07.007.

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