

## Case Report

# Recurrent Episodes of ST-Elevation Myocardial Infarction in a 19-Year-Old Male with Fontan Circulation

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## Abstract

We describe a case of a 19-year-old male with history of single ventricle physiology status post-Fontan procedure at the age of two who developed thromboembolic phenomena involving his splenic, renal and coronary arteries resulting in multiple infarcts and recurrent in-hospital acute ST-Segment Elevation Myocardial Infarction (STEMI) treated by emergent Percutaneous Coronary Intervention (PCI). This case highlights multiple aspects and challenges of managing young patients with congenital heart disease.

**Keywords:** Congenital heart disease; Heart failure; Thromboembolism; Anticoagulation; Acute coronary syndrome

## Abbreviations

ECG: Electrocardiogram; LAD: Left Anterior Descending Artery; STEMI: ST-Segment Elevation Myocardial Infarction; DOAC: Direct Oral Anticoagulants; CVICU: Cardiovascular Intensive Care Unit

## Case Presentation

19-year-old male with a history of single ventricle physiology (S,L,L) including L-transposition of the great arteries, pulmonary valve atresia, left atrioventricular valve atresia, large unrestrictive VSD, unrestrictive secundum ASD, and left pulmonary artery stenosis status post staged corrective heart surgery with eventually extracardiac Fontan at the age of two who presented to our emergency department complaining of abdominal pain for five days, associated with nausea and vomiting. Prior to his presentation to our facility, he was recently discharged from a local hospital after evaluation of chest pain with left sided weakness; he was found to have severe systolic and diastolic dysfunction, and age indeterminate thromboembolic stroke. His cardiac catheterization during this admission did not show any coronary vasculature abnormalities. He was discharged on aspirin, furosemide and rivaroxaban due to patient's refusal to be treated with warfarin. His medications history also includes daily prophylactic aspirin since his Fontan procedure, but he reported not taking it.

On exam he was vitally stable with left upper quadrant tenderness. Labs notable for normal complete blood count and kidney function. B-type natriuretic peptide resulted 1235pg/mL (normal range: ≤100pg/mL). Chest radiograph showed evidence of pulmonary edema and CT abdomen/pelvis revealed acute splenic infarct and left renal infarct. He was admitted to the Cardiovascular Intensive Care Unit (CVICU) for management of acute exacerbation of heart failure, and acute infarcts involving the spleen and left renal arteries. He was started on infusions of milrinone and furosemide; as well as heparin infusion for management of heart failure and thromboemboli respectively. Our adult congenital heart disease and hematology teams were also involved in his inpatient care.



**Figure 1:** Electrocardiogram. Electrocardiogram depicting anterolateral ST-Segment Elevation Myocardial Infarction (STEMI).

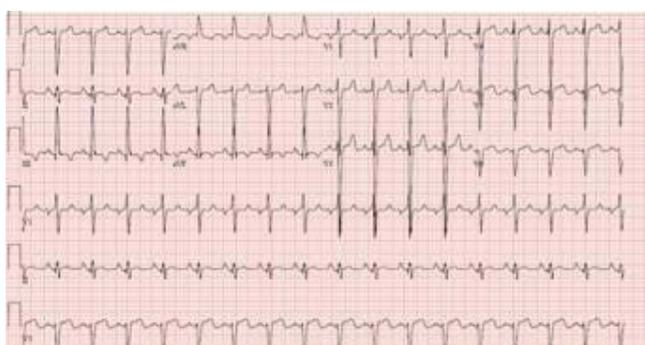


**Figure 2:** Coronary angiogram. Angiography of the morphologic left coronary artery demonstrating a cutoff in the mid Left Anterior Descending artery (LAD).

Shortly following admission, he reported acute severe non-radiating substernal pressure-like chest pain. Clinical examination was notable for tachycardia. Electrocardiogram (ECG) showed



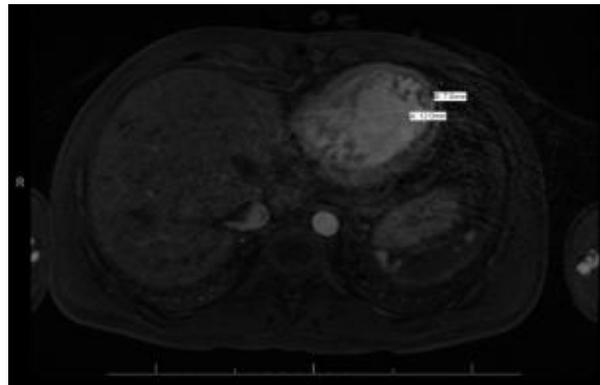
**Figure 3:** Coronary angiogram. Angiography post thrombectomy with blood flow reestablished to the Left Anterior Descending Artery (LAD); the terminal vessel remained occluded.



**Figure 4:** Electrocardiogram. Electrocardiogram showing resolution of ST-segment elevations in the anterolateral leads following thrombectomy.

ST elevation in the anterolateral leads (Figure 1). Patient was immediately taken to the cardiac catheterization laboratory where coronary angiogram showed situs inversus, and the morphologic left coronary artery demonstrated a thrombotic occlusion in the mid Left Anterior Descending Artery (LAD) (Figure 2). A thrombectomy was performed and coronary flow was re-established; however, this intervention did not restore distal flow. Stenting was not performed given the embolic nature of the lesion. The final angiography shot showed TIMI3 flow in all vessels with persistent limited flow and occlusion in the distal LAD (Figure 3). The patient was transferred back to the CVICU and continued on antiplatelet therapy, with resolution of symptoms and ECG changes (Figure 4). Troponin peaked at 16.5ng/mL (normal range:  $\leq 0.03$ ng/mL).

Following clinical stabilization, a cardiac MRI performed with contrast showed globular, hypointense filling defect at the apex of the systemic ventricle (1.2 cm x 0.8 cm), consistent with thrombus (Figure 5). Additional work up was unrevealing (including causes for a hypercoagulable state). He was eventually transferred out of the CVICU to the telemetry cardiology ward in a stable condition. Four days following the initial ST-Segment Elevation Myocardial Infarction (STEMI) episode, he again developed acute chest pain and ECG showed lateral STEMI. However, a repeat coronary angiogram demonstrated no change and subsequently the ECG normalized. Given the patient's refusal to be treated with warfarin, he was



**Figure 5:** Cardiac Magnetic Resonance Imaging (MRI). Cardiac MRI demonstrating globular, hypointense structure at the apex of the systemic ventricle (1.2 cm x 0.8 cm) consistent with thrombus.

eventually transitioned to Direct Oral Anticoagulant (DOAC) and discharged home, as he remained chest pain free and stable.

## Discussion

Fontan procedure is the final stage of surgical palliative approach to manage patients with single ventricle physiology, it is usually performed at 18 months to 4 years of age [1]. Following this procedure, multiple complications have been reported including but not limited to hepatic dysfunction, myocardial dysfunction, atrial rhythm disturbances, worsening cyanosis and thromboembolic events [2]. Thromboembolic events in particular impose high risk of morbidity and mortality with an incidence ranging from 3-20 % [3]. The incidence of intracardiac thrombus remains unknown given the presence of asymptomatic cases, however it is reported as high as 33% [4]. There are only few reported cases of myocardial infarction as a result of thromboembolism in patients with Fontan circulation [5].

The efficacy of primary prophylaxis of thromboembolic events in Fontan circulation patients remains a debatable issue. Lower death rates have been reported in patients with Fontan circulation who are anticoagulated or in whom antiplatelet therapy is used [6]. Studies showed no superiority when using anticoagulation with warfarin compared to antiplatelet, therefore current guidelines recommend using aspirin for all patients with history of Fontan procedure [1]. According to the 2018 AHA/ACC guidelines for the management of adults with congenital heart disease, current indications for anticoagulation with warfarin in patients with Fontan circulation include cases with atrial arrhythmias, residual atrial septal defects/fenestrations or thromboembolic event [6].

The guidelines however do not specifically recommend the use of DOAC at present time given the scarcity of evidence in this area. Despite that, the published literature supports the safety and non-inferiority of DOAC when compared to warfarin in Fontan circulation patients [7-11].

Fortunately, the occurrence of STEMI following thrombus formation is extremely rare, however among reported cases, primary angiography with balloon dilation and thrombectomy is the main approach for treatment [5]. Our patient was taken to the catheterization laboratory following each STEMI episode and treated

with thrombectomy following the first episode. On both occasions he was treated with aspirin, heparin and tirofiban infusion for 18 hours. These interventions resulted in resolution of symptoms. He was finally discharged home on Aspirin and DOAC.

## Conclusion

In conclusion, further research is needed to investigate the role of DOAC in treatment and prevention of thromboembolic events in patients with Fontan circulation. Our case specifically drew attention to patient's history of non-adherence to therapy as it could play major role in the decision-making regarding anticoagulation.

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