



Embolic Myocardial Infarction in a Patient With a Fontan Circulation

World Journal for Pediatric and
Congenital Heart Surgery
2014, Vol. 5(4) 631-634
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DOI: 10.1177/2150135114540180
pch.sagepub.com



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Abstract

Coronary artery embolism is an uncommon cause of acute myocardial infarction (MI). We present a patient with pulmonary atresia and severe right heart hypoplasia who underwent a lateral tunnel Fontan procedure in childhood and presented with an acute ST-segment elevation MI at 19 years of age. In addition to the known risk of thrombotic complications associated with a Fontan circulation, potential predisposing factors to thromboembolism in this patient included a right ventricle to left anterior descending coronary connection and a Fontan baffle leak. The patient was treated with device closure of the baffle leak and anticoagulation. This is one of the first reports of an embolic MI in a patient with a Fontan circulation. The optimal method of reducing thromboembolic risk in this patient, and those with a Fontan circulation in general, is complicated and no consensus exists.

Keywords

Fontan, myocardial infarction, embolism, tricuspid atresia

Submitted January 23, 2014; Accepted May 20, 2014.

Introduction

Coronary artery embolism is a well-known cause of acute myocardial infarction (MI). Although there are reports of paradoxical embolism through a patent foramen ovale, there are few reports of MI secondary to thromboemboli from other sources. We describe a young adult with a Fontan circulation who presented with a thromboembolic MI. Our patient consented to the publication of this case report.

Case Report

The patient is a 19-year-old woman born with pulmonary atresia, an intact ventricular septum, normally related great arteries, severe tricuspid valve and right ventricular (RV) hypoplasia, and RV to coronary artery connections without an RV-dependent coronary circulation. At two days of age, a 5 mm left modified Blalock-Taussig shunt was performed. Four months later, she underwent surgical atrial septectomy after an unsuccessful attempt at balloon septostomy for atrial septal restriction. At three years of age, an intracardiac lateral tunnel Fontan procedure was performed, including a bidirectional Glenn anastomosis and left pulmonary artery augmentation due to stenosis at the site of the shunt. Pulmonary valve atresia was confirmed when the main pulmonary artery was divided and oversewn.

The patient did well on chronic aspirin therapy until the day of admission, when she developed substernal chest pressure at

rest. She presented to a local emergency department where inferior ST-segment elevation was noted on her electrocardiogram. Heparin, aspirin, and clopidogrel were initiated, and she was transferred to our institution. Emergent left heart catheterization revealed an ectatic left anterior descending (LAD) coronary artery with a fistula to the RV apex, a small but normal left circumflex artery, a discrete filling defect in the right coronary artery (RCA), and no RV to coronary connections other than the LAD fistula (Figure 1). Thrombectomy and balloon angioplasty were performed with restoration of a normal RCA lumen.

She had normal hemoglobin, platelets, prothrombin time, renal function, and an initial negative troponin I that rose to 25 ng/dL (normal <0.01 ng/dL) after catheterization. Her oxygenation was 96% on room air, and she was normotensive. She had no evidence of heart failure and a peripheral vascular examination was normal.

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Abbreviations and acronyms

LAD	left anterior descending
MI	myocardial infarction
MTHFR	methylenetetrahydrofolate reductase
RCA	right coronary artery
RV	right ventricular

She did well after catheterization, without heart failure or arrhythmias. A lower extremity venous ultrasound failed to show any deep vein thrombosis. Cardiac magnetic resonance imaging showed evidence of prior infarction in the LAD territory, an acute infarction in the RCA territory, a Fontan baffle leak, drainage of the coronary sinus to the morphologic right atrium on the pulmonary venous side of the Fontan baffle, left subclavian vein occlusion with left arm veins draining to the left pulmonary vein via collaterals, and a fistula between the LAD and the RV (Figure 2). Transesophageal echocardiography did not reveal any evidence of intracardiac thrombus. A bubble study with saline injection in a left arm vein was strongly positive for right to left shunting. A workup for thrombophilic disorders was unrevealing, including normal prothrombin, partial thromboplastin, thrombin time, fibrinogen level, protein C and S function, antithrombin function; no factor V Leiden, prothrombin, or methylenetetrahydrofolate reductase (MTHFR) mutations; and no evidence of anticardiolipin or β -2 glycoprotein antibodies.

She was discharged on warfarin and aspirin and remained free from further events. An exercise stress test revealed reduced exercise capacity, no arrhythmias, and reduction in the arterial oxygen saturation to 88%. Approximately 18 months after the MI, she underwent repeat cardiac catheterization, at which time the Fontan circulation and RV to LAD connection were evaluated more closely. During access, it was confirmed that there was occlusion of the right and left femoral and left subclavian veins, and the left arm circulation returned to the heart via venous collaterals. There was a complex, medium-sized leak in the mid-anterior portion of the baffle, with right to left shunting of contrast. This was closed with a 6-mm Amplatzer vascular plug II device (St Jude Medical, Minneapolis, Minnesota). There was a trivial amount of antegrade flow through the tricuspid valve. The LAD to RV fistula was nonrestrictive, and although the LAD was uniformly dilated, there was a relatively small amount of flow into the RV. The only outflow from the RV was the fistula itself, and there was retrograde systolic flow in the LAD. After discussion, it was elected not to attempt device closure of the fistula.

Discussion

Thromboembolic complications are relatively common in individuals with a Fontan circulation, and although systemic embolization causing cerebrovascular events has been reported after a Fontan procedure, embolic MI is rare.¹⁻³ Although reports of emboli originating from the left heart⁴ or paradoxical embolism through a patent foramen ovale⁵ have been reported to cause MI, this is one of the first descriptions of MI caused by embolism in a patient with complex congenital heart disease with multiple potential embolic sources.

Given the appearance of a discrete filling defect with a normal underlying RCA on cardiac catheterization, along with no procoagulant disorder, we determined that the most likely diagnosis in this patient was a coronary artery embolism. Her workup revealed several potential sources of thromboembolism, although none were the obvious culprit. There were right to left communications through a Fontan baffle leak and venous collaterals from the upper extremity veins to the pulmonary veins. In addition, she had a large ectatic LAD with a fistula to the hypoplastic and highly trabeculated RV, which had almost no inflow through the hypoplastic tricuspid valve, and the only outflow from the RV was the fistula. Either the dilated LAD or the RV may have been predisposed to thrombus formation, which could have propagated retrograde as a result of systolic retrograde flow. She also had multiple central venous occlusions, although there was no evidence of associated acute thrombosis on imaging.

In Fontan patients generally, and this young patient in particular, there are a number of factors that may increase the risk of thromboembolism.^{1,6} In addition to potential mural stasis in the Fontan pathway in the presence of prosthetic material, the coagulation cascade and platelet function are often abnormal in these patients.¹ Systemic venous hypertension can contribute to congestive hepatopathy, leading to further deficits in clotting factors.^{1,6} Various other factors may also predispose to thrombosis, including venous insufficiency, various causes of right to left shunting, atrial arrhythmias, intra-atrial synthetic material, and others. Patients with pulmonary atresia and intact ventricular septum frequently have coronary artery to RV connections, and in some cases coronary steal or stenosis can lead to MI,⁷ but we are not aware of reports of coronary thromboembolism in this population.

There is no clear consensus regarding medical treatment to prevent thromboembolic events in patients after a Fontan procedure.^{1,2} Given the suspected systemic embolic event, dual treatment with warfarin and aspirin was initiated. Closure of the baffle leak at a subsequent catheterization seemed appropriate and low risk; however, there remained right to left shunting through the profuse left arm venous collaterals draining to the pulmonary venous system, which were multiple and impractical to close. A more complex dilemma was whether to close the RV to LAD fistula in the catheterization laboratory. The patient had evidence of prior LAD infarction, and thrombus formation was a possibility in the ectatic LAD or the hypoplastic RV. In contrast to most large coronary artery fistulas, there was no other outflow from the RV in this patient so the volume of shunting through the fistula was quite small; thus, the indication for closure would have been primarily to reduce the risk of thromboembolic events to which the fistula may have been unrelated. This decision was complicated by recent data demonstrating a substantial risk of MI or coronary thrombosis after closure of coronary artery fistulas.⁸ All things considered, the decision was made to treat the LAD fistula with medical therapy alone with close follow-up. On follow-up one year later, she had no further evidence of thromboembolic events on combined aspirin and warfarin therapy.

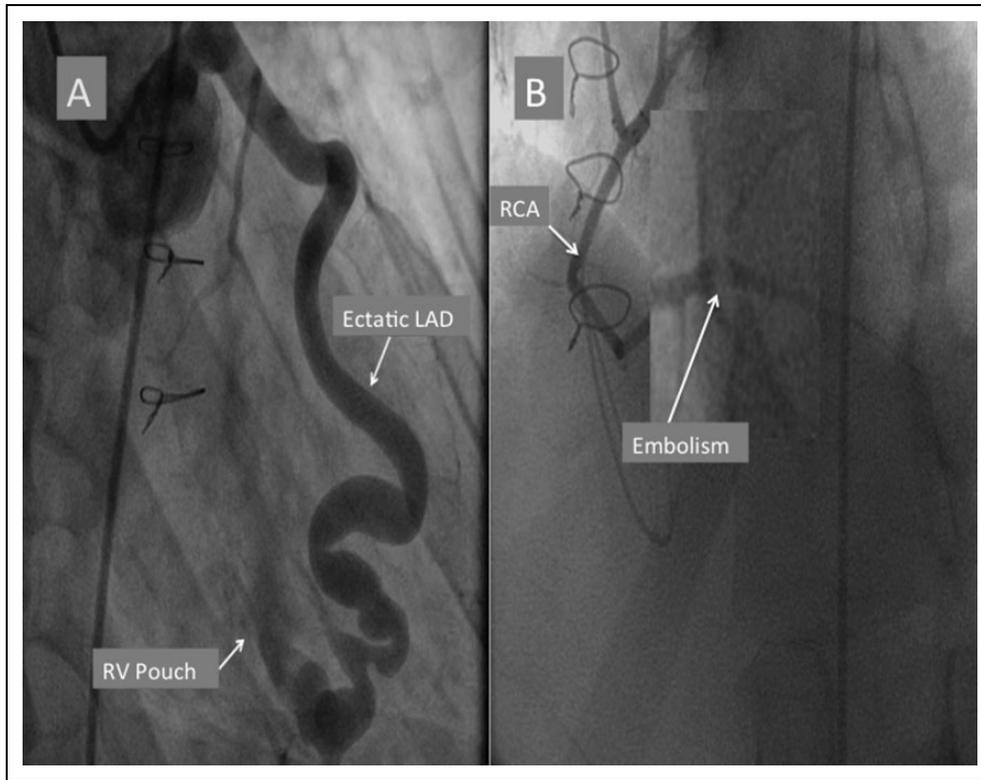


Figure 1. (A) Coronary angiography showing an ectatic left anterior descending artery (LAD) with a fistula to the right ventricle (RV), and (B) the acute right coronary artery (RCA) embolism.

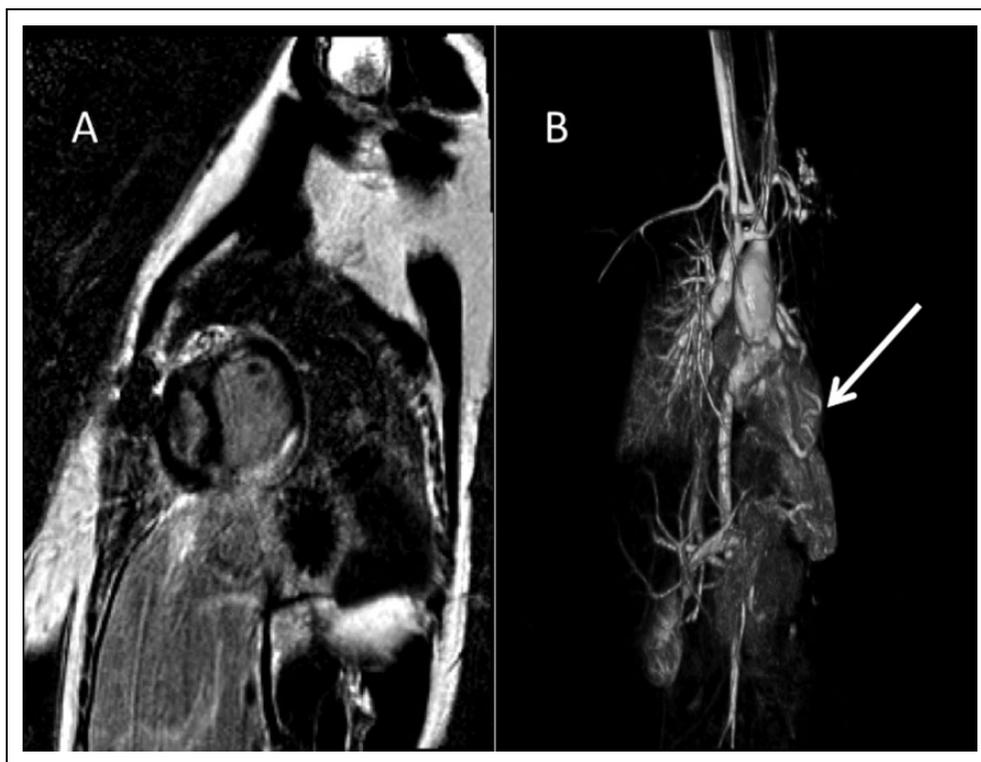


Figure 2. A, Cardiac magnetic resonance imaging (MRI) demonstrating late gadolinium enhancement in the mid-inferior wall (transmural) and also a small (<50%) area in the mid-anterior left ventricular wall suggesting prior myocardial infarction. B, Large ectatic LAD to RV pouch fistula (arrow).

This is one of the first described cases of MI secondary to systemic embolization in a patient with a Fontan circulation. There is no clear consensus on therapy to prevent thrombosis after a Fontan procedure, as long-term randomized trials would be difficult to perform.² Instead, the clinician must weigh the patient-specific risk against the risk of lifelong anticoagulation. Although new imaging modalities can localize sources of shunting in patients with complex congenital heart disease, there is no evidence to guide the closure of these connections in patients with or without history of embolism, and in some cases, such as diffuse systemic to pulmonary venous collaterals, complete closure may be very difficult.

Declaration of Conflicting Interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

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