

Title: Detection of Occult Thrombosis in Individuals with Fontan Circulation

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Background: In patients with Fontan circulation, transesophageal echocardiogram (TEE) studies have reported prevalence of intracardiac thrombosis between 17% to 33% with a reported mortality of 25% with Fontan-associated thromboembolism. However, prior studies have been confounded by a lack of universal TEE screening, or TEE imaging used primarily for patients presenting with events suspicious for thromboembolism. Delayed enhancement cardiac magnetic resonance (DE-CMR) with long TI is superior to transthoracic echo and TEE for detection of intracardiac thrombus in adult studies. Our objective was to determine prevalence of occult cardiac thrombosis in patients undergoing clinically indicated CMR.

Methods: A retrospective chart review was performed of individuals with Fontan circulation who underwent DE-CMR imaging at Duke University Hospital from October 28, 2008 through February 28, 2020. Demographics were reviewed including anthropometric data, medical and surgical history, imaging results, and outcomes data (thromboembolic complications, subsequent imaging results, need for further interventions, and death). DE-CMR was performed using post-contrast, long TI imaging (600-800 msec) to null avascular tissue such as thrombus. All study images were reviewed for adequacy and agreement with clinical report.

Results: A total of 121 unique individuals were identified meeting inclusion criteria with a total of 183 scans. The median age at time of the CMR was 18 years (range 2-50 years) with 43% female and 57% male individuals. The median age at Fontan procedure was 3 years (range 1-25 years). Type of Fontan procedure is noted in Figure 1. Of 183 studies, only one demonstrated a Fontan circuit thrombus (0.5% of studies). This patient was 32 years old at time of imaging and had a fenestrated lateral tunnel Fontan at age 23. Given concern for a paradoxical embolic myocardial ischemic event, transthoracic echocardiogram was initially performed and did not identify a thrombus. Ongoing clinical concern prompted CMR revealing small thrombus in the Fontan circuit as well as the right atrial appendage. Diagnostic and interventional catheter-based study confirmed thrombus burden.

Conclusions: Despite previous echocardiographic reports of high prevalence of occult thrombosis in patients with Fontan circulation, we found very low prevalence using DE-CMR. With expected results from the UNIVERSE study and the added benefits of volumetric and functional assessment, interval DE-CMR along with echocardiography should be considered as part of routine surveillance of individuals with Fontan circulations.

Types of Fontan Procedures

