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LATE THROMBOSIS OF THE NATIVE AORTIC ROOT AFTER FONTAN PROCEDURE FOR HYPOPLASTIC LEFT HEART SYNDROME

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Introduction

Thrombus formation within the native aortic root after Norwood palliation for hypoplastic left heart syndrome (HLHS) is considered to be a potential lethal condition. In particular, in aortic atresia context, the ascending aorta has the role of supplying the coronary circulation. We describe a case of thrombosis into the ascending aorta following a Fontan palliation for HLHS.

Case report

A 16-year-old female with HLHS and aortic atresia was initially palliated with stage I Norwood procedure, at 9 months of age with bidirectional Glenn and at 5 years of age with fenestrated extracardiac Fontan (ECF). She was on aspirin. Eleven years later, she experienced an out-of-hospital cardiac arrest. Transthoracic echocardiogram (TTE) showed new onset of severe single ventricle dysfunction. The patient underwent a Cardiac CT scan (Figure 1). We proceeded with systemic thrombolysis with a consequent improvement in the clinical condition. In the following days, we performed an transesophageal echocardiogram (TEE) (Figure 2) and serial CT scan (Figure 3). Heparin was initiated and subsequently imbricated with warfarin. The patient was discharged home with a resolution of the native aortic root thrombosis (NART), a depressed systemic ventricular function and a dual therapy with an anticoagulant (warfarin) and an antiplatelet (aspirin) drug.

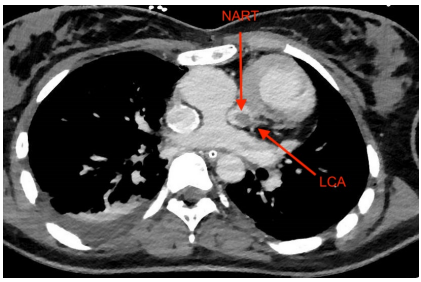

Figure 1. Cardiac CT scan showed a 13x13mm native aortic root thrombosis (NART) adherent to the left coronary sinus of Valsalva and extended to the ostium of the left coronary artery (LCA) which was totally occluded, the right coronary artery (RCA) appeared unobstructed.

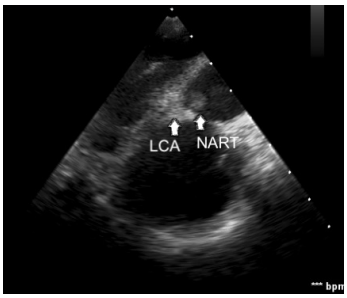

Figure 2. The transesophageal echocardiogram (TEE) showed a slight reduction of the native aortic root thrombosis (NART).

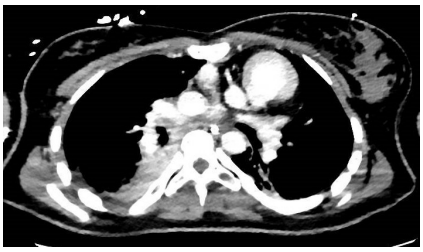

Figure 3. Serial cardiac CT scan revealed a progressive reduction of the thrombus, with a complete recanalization of the left coronary artery (LCA).

Discussion and conclusion

NART after the ECF operation in HLHS is very uncommon. Hemodynamic changes appear to be the dominant etiologic factor for the NART: in patients with HLHS anatomy, the trivial or absent antegrade flow through the native aorta may cause stasis of blood at the level of the aortic root, potentially predisposing to a slow blood swirl in the native aortic root with consequent thrombus formation. The hypercoagulable state of patients with single-ventricle may also contribute. In case of new ventricular arrhythmias or evidence of myocardial ischemia (MI) in patients after Norwood palliation, there should be a high suspicion for NART that should be sought through TTE, TEE, CT or cardiac catheterization. Currently, the management of patients with NART can include lone anticoagulation, systemic thrombolysis, directed thrombolysis injected in the native aortic root and surgical thromboembolectomy and must be customized considering risks and benefits of each individual procedure. Patients with atretic aortic valve and a relatively long and thin native aorta may benefit from lifelong anticoagulation to prevent NART.