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Native Aortic Root Thrombosis in Hypoplastic Left Heart Syndrome: An Unusual Case Soon After Atrial Septal Stenting and Literature Review with an Outlook to Diagnosis and Management

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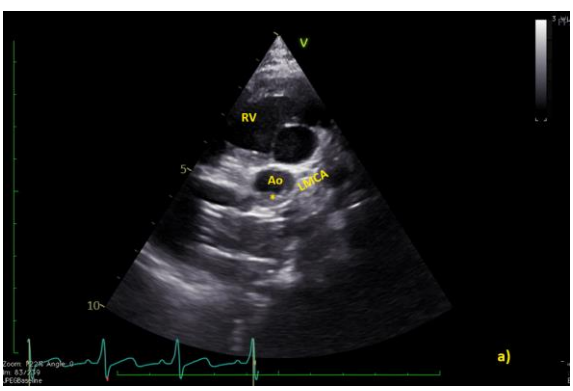
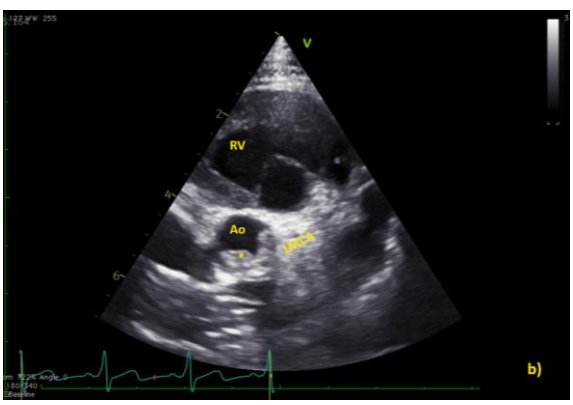
We reviewed incidence, diagnosis, and management of native aortic thrombosis in HLHS after different stages of Fontan palliation through a systematic literature search. Age at diagnosis ranged from 13 days to 18 years. Clinical presentation varied from incidental **findings, chest pain and/or electrocardiographic abnormalities, cardiac arrest, and transient ischemic attack**. Diagnosis was feasible in most of the cases with only transthoracic echocardiography. Mostly (59.4%) were treated with anticoagulation, while others with surgical (18.7%), direct (12.5%) or systemic (9.3%) thrombolysis.

Patient: 3 years old male child with hypoplastic left heart syndrome (HLHS) and severely hypoplastic but patent mitral and aortic valves after Norwood-Sano palliation, status post left heart decompression by percutaneous stenting of the atrial septum. The diagnosis of thrombus formation in the native aorta was incidental, with a child completely asymptomatic and progressively subsided in a few days with heparin infusion and chronic warfarin therapy.

We retain that this complication was not related to procedure itself, rather to a change in the hemodynamic which diminished the anterograde flow to the aorta.





Native aortic thrombosis in HLHS may occur at different ages, with a wide spectrum of presentation, from incidental finding to sudden major event. Diagnosis **is feasible with transthoracic echocardiography** and management with anticoagulation is effective, despite the incidence of major events remaining high.

The increased awareness of this rare complication has led to an increasing diagnostic rate in the last few years, with multiple cases described. This remarks the importance of systematically echocardiographic investigation of the native aorta as a potential site for thrombus formation in patients with HLHS and hypoplastic inflow and outflow pathways, even when hemodynamic and clinical conditions seem to be optimal.