Aortic root thrombosis with coronary embolization following neo-aortic reconstruction in a child with hypoplastic left heart syndrome

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Abstract

In the recent era, the diagnosis, treatment options, postoperative management and outcomes of infants born with hypoplastic left heart syndrome (HLHS) have undergone dramatic changes. As is the case with many other novel treatment modalities used for congenital heart diseases, data concerning the long-term outcomes and complications of the various strategies become gradually more available as the numbers of survivors grow. In general, complications of the three-stage surgical palliation used for HLHS tend to occur most commonly following the first-stage surgery. Post-stage 2 complications are substantially less common, and centre on the procedure itself and the unique physiology of the cavopulmonary connection. In the following case report, we describe a relatively rare adverse outcome that occurred following a stage 2 surgery in the form of native aortic root thrombosis extending to the coronary arteries. The selected methods of treatment used in the catheterization laboratory and later in the operating theatre, as well as its outcomes are described.

Keywords: Hypoplastic left heart syndrome • Thrombosis • Thrombolysis

CASE REPORT

The patient was a 13-month-old female born with hypoplastic left heart syndrome (HLHS), severe mitral stenosis and a nearly-atretic aortic valve with a z-score of −5.8 and minimal antegrade flow, who initially underwent a hybrid stage 1 procedure (patent ductus arteriosus stent, branch pulmonary artery (PA) banding and balloon atrial septostomy), followed by comprehensive stage 2 procedure (PA band removal, Damus–Kaye–Stansel (DKS) type aortic reconstruction, Glenn anastomosis and atrial septectomy) at 4 months of age. Following the DKS anastomosis, the length of the native ascending aorta was 1.7 cm. The diameter at the sinuses was 7.5 mm and at the sino-tubular junction 5 mm, both yielding a z-score of −3.5. Per institutional protocol, she was treated for 6 weeks with low-molecular-weight heparin for thromboprophylaxis following her stage 2 procedure.

Nine months after surgery, she was brought by her parents to an external facility due to increasing fatigue on exertion, agitation and an unexplained syncopal episode. Electrocardiogram and transthoracic echocardiogram revealed new complete atroio-ventricular block with mean ventricular escape rate of 40 beats per minute and new moderate-to-severe systemic right ventricular (RV) dysfunction, respectively. In addition, a mobile echogenic mass suspected to be an intracavitary thrombus was present in the hypoplastic left ventricle (Fig. 1). Dobutamine and isoproterenol infusions were started, and she was transferred to our institution for further management. Upon arrival, sinus rhythm was regained and initial laboratory studies showed troponin I values of ~150 times the upper limit values. An urgent cardiac magnetic resonance imaging study was obtained, demonstrating a dilated, severely dysfunctional RV with hypokinesis of the posterior and free wall. Delayed subendocardial Gadolinium enhancement in the hypokinetic regions of the RV was noted, as was the previously recognized LV thrombus. Given the elevated troponin levels and the delayed Gadolinium enhancement, an ischaemic aetiology was strongly suspected, and the patient was transferred to the catheterization laboratory for further investigation. During the catheterization, a gentle hand injection into the native hypoplastic ascending aorta revealed a large, non-calcified thrombotic mass in the native aortic root, as well as filling defects in the left main and the circumflex coronary arteries which appeared to branch in a normal anatomic fashion (Fig. 1B). After careful consideration of all treatment options, we proceeded with catheter-directed thrombolytic therapy into the native aortic root under continuous monitoring in the catheterization laboratory. A bolus administration of tissue plasminogen activator was thus delivered, followed by a repeat slow hand contrast injection. This showed a slight reduction in the thrombotic mass in the aortic root and left main coronary artery, but also a new cut-off defect of the first diagonal artery (Fig. 1C), likely as a result of thrombus lysis and embolization. As this was not accompanied by ST-segment changes, haemodynamic changes or arrhythmias, the patient was transferred to the intensive cardiac care unit, where systemic thrombolysis was administered overnight. The following day planned repeat angiography was obtained, which revealed a substantial reduction of the thrombotic
mass in the aortic root. Despite a slow hand contrast injection, additional thrombus dislodgement and embolization occurred, this time down the right coronary artery (Video 1). The embolus appeared to have undergone immediate lysis without blocking either the main or any of the branch vessels, as repeat contrast angiography showed grade 3 thrombolysis in myocardial infarction coronary flow. Once again, this was not accompanied by haemodynamic, ST-segment changes, or arrhythmias. Thrombolytic therapy was continued for additional 24 h in the intensive care unit.

Her further hospitalization course was not favourable; Anticoagulation therapy was commenced, initially with heparin and later warfarin. Although she remained stable with this management and within therapeutic international normalized ratio (INR) ranges, serial echocardiograms showed recurrent thrombus progression in the native aortic root and worsening RV function, which was accompanied by a cerebrovascular embolic event.

Given this apparent lack of response to anticoagulation and the risk of further thromboembolic events, the valve was sawn closed (Fig. 2A–C). The patient survived the procedure, yet her ejection fraction remained low and she had persistent heart failure symptoms. She was subsequently listed for a heart transplantation and currently awaits on the transplant list.

**DISCUSSION**

Following neo-aortic reconstruction in children born with HLHS, the short, often diminutive residual root segment of the native aorta remains a conduit for retrograde blood flow to the coronary vessels arising from it. Although the occurrence of native root thrombosis has been rarely reported, one post-mortem study reported that among 122 patients who had undergone the Norwood procedure, the incidence of coronary insufficiency was as high as 27%, making it the most important cause of death in this series [1]. Based on several prior reports [2–4], common manifestations of root thrombosis in single ventricle patients include conduction disturbances, congestive heart failure and thromboembolism, including to the cerebral vasculature. In the absence of large cohort data, hypothesized risk factors include anatomic native aortic valve abnormality prompting stasis, prolonged hospitalizations, absence of anticoagulant/anti-aggregant therapies and surgery-related issues, such as the length of the remnant native root (longer remnant may increase the risk for thrombus formation) and the amount of sutures placed [2–4]. In addition, several small cohort studies demonstrated that some patients may obtain the tendency for hypercoagulability [5], likely as a result of the altered haemodynamic state following these procedures, but possibly also of congenital coagulopathies. Data on the risks and benefits of possible therapeutic interventions is absent, leaving intervention-related decisions to be made on a case-by-case basis and based on the specific clinical scenario. Options include anticoagulation alone, systemic thrombolysis, directed thrombolysis injected to the aortic root and surgical thromboembolectomy.
Meticulous clinical and imaging-based follow-up is recommended for the single-ventricle post stage 2 patients. The optimal period for anticoagulation administration and whether certain sub-populations can benefit from longer periods are issues that remain to be addressed in larger studies. In addition, data from large patient cohorts on the long-term outcomes and adverse events after the stage 2 procedure would be an important contribution to the paediatric cardiology literature.

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REFERENCES