Coronary Thrombo-embolism in Hypoplastic Left Heart Syndrome - A treatable cause of interstage mortality

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Introduction
Mortality rates in hypoplastic left heart syndrome (HLHS) continue to improve since the introduction of staged surgical palliation. However, inter-stage deaths continue to be challenging to prevent. This case presents a child with HLHS post Glenn shunt presenting with acute ST-elevation myocardial infarction (MI). He was treated using a well established procedure in treatment of acute MI in adults. Treatment of coronary thrombo-embolism in the setting of uni-ventricular circulations has not been previously reported. This case demonstrates the importance of any evidence of myocardial ischaemia in these patients and presents a novel and potentially lifesaving therapy for consideration.

Methods
A three-year old boy with HLHS post Glenn shunt presented with sudden onset, classical symptoms and ECG changes consistent with acute ST-elevation MI. Echocardiography showed acute deterioration in right ventricular systolic function. He was initially treated with thrombolysis and inotropic support for 24 hours but inferior ST-elevation persisted with development of Q-waves and inotropic dependence. He was therefore taken to the catheter laboratory. Angiography showed a dominant right coronary artery with almost total occlusion of the distal right main coronary artery. Using an Export AP aspiration catheter (Medtronic, Minneapolis MN), a large thrombus was aspirated and repeat angiography showed complete thrombus resolution with normal vessel patency. There were no procedural complications.

Results
There was an immediate improvement in ventricular function post-procedure. He was commenced on beta-blocker and ACE inhibitor and was discharged home approximately three weeks post-intervention. He has been seen at one and six weeks and 3 months following discharge and his echocardiogram shows ongoing functional improvement.

Conclusions
This case illustrates the novel application of a proven adult coronary intervention in complex paediatric congenital heart disease. This has not been reported previously. The importance of this intervention is because the entire circulation is dependent on an essentially single coronary artery system. The effects of acute occlusion therefore cannot be mitigated by collateralisation leading to more profound circulatory effects. Furthermore, it suggests one possible aetiology of inter-stage deaths. Finally, the case demonstrates the additional benefits of collaboration between adult and congenital cardiologists providing alternative approaches for challenging situations.