Case report

Veno-Venous Coiling as A Strategy to Wean Extracorporeal Membrane Oxygenator Post Adult Fontan Completion

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Abstract:

We discuss a case of veno-venous collaterals in a patient with Fontan circulation post complex redo-surgery, which impeded veno-venous extracorporeal membrane oxygenation (VV-ECMO) weaning.

A 31-year-old male, born with complex congenital heart disease (D-transposition of the great vessels, tricuspid atresia, aortic coarctation, ventricular septal defect, and hypoplastic right ventricle) with single ventricle physiology. The patient underwent multiple cardiac surgeries leading to Fontan completion. He subsequently developed significant aortic regurgitation with progressive left ventricular dilatation requiring multiple readmissions for congestive heart failure. Despite maximal medical optimisation he remained symptomatic and was offered further surgery. Complex 6th redo-operation with aortic valve replacement, modification to intracardiac fenestrated Fontan and insertion of intra-aortic balloon pump was performed. At 72 hours postoperative, VV-ECMO was required for refractory hypoxia. Right heart catheterisation was performed with coiling of two large fistulous venous connections. Post-coiling saturations improved from 92% to 98%. The patient was successfully weaned from VV-ECMO and decannulated the following day. This case demonstrates successful embolisation for veno-venous collaterals causing significant right to left shunt and hypoxia, in a patient with Fontan circulation.

Key words: fontan circulation; venous collaterals; coiling; extracorporeal membrane oxygenation; ECMO; ACHD; adult congenital heart disease

Abbreviations:

ACHD – Adult Congenital Heart Disease

LVEDD - Left ventricular end diastolic volume

VV-ECMO - veno-venous extracorporeal membrane oxygenation

Introduction

The Fontan procedure, a palliation procedure, has been successfully performed for over 30 years. These patients are now living into adulthood and developing long-term complications of single ventricle driven physiology. Veno-venous collateral formation is a known complication, with debated aetiology. The current theory is that decompressing collaterals form in response to systemic venous hypertension (1-3). The benefit of coiling chronic collateral vessels provides no advantage to these patients (1). We discuss a patient with significant venous collaterals identified postoperatively causing refractory hypoxia.

Case Presentation:

A 31-year-old male, presented with heart failure. He was born with complex congenital heart disease and single ventricle physiology (D-transposition of the great vessels, tricuspid atresia, aortic coarctation, ventricular septal defect, and hypoplastic right ventricle). The patient had multiple cardiac surgeries as a child (two coarctation repairs, Damus-Kaye-Stansel procedure and Glenn shunt) leading to Fontan completion. He progressively developed severe symptomatic aortic regurgitation, left ventricular dilation and heart failure requiring multiple readmissions. A holter monitor demonstrated episodic atrial tachycardia. Preoperative CT demonstrated no collateral vessels and right heart catheter showed normal pulmonary pressure and no venous collaterals. Transthoracic echocardiogram demonstrated dilated left ventricle (LVEDD 70mm) EF 47%, neo-aortic valve with 3-4/4 regurgitation, 2-3/4 mitral regurgitation, hypoplastic right ventricle with tricuspid atresia, and neo-pulmonary

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valve 2/4 regurgitation. Despite medical optimisation he remained symptomatic. Redo operation was performed with 6th sternotomy, neoaortic valve replacement, central Alfieri type plication of the neopulmonary valve, modification to intracardiac fenestrated Fontan and insertion of intra-aortic balloon pump. The patient returned to intensive care on modest inotropes and acceptable haemodynamics.

At 72 hours postoperative, VV-ECMO was required for worsening refractory hypoxia. Despite VV-ECMO flow of 3.33L, sweep gas flow of 5L/min, Fi02 70 and ventilator settings of Fi02 60, PEEP 7cmH2O and tidal volumes of 430ml, saturations remained at 91%. Blood gas demonstrated pH 7.51, O2 62mmHg and pCO2 35mmHg. Postoperative CT demonstrated venous collaterals, and a right heart catheter was

performed to confirm. The tight heart catheterisation demonstrating two large fistulous venous connections from the innominate vein to the right and left superior pulmonary veins (Figure1A, B) with significant right to left shunt Qp: Qs 1.5:1. Following multidisciplinary discussion, the two collateral vessels were coiled (Figure1C, D) with multiple retrievable Boston Scientific Interlock TM coils 3 x (6mm x 10cm), 2 x (6mm x 20cm), 8mm x 20cm, 10mm x 30cm and 2 x (10mm x 50cm). Saturations rapidly improved from 92% to 98% and arterial blood gas demonstrated pH 7.41, pO2 69mmHg and pCO2 38mmHg. VV-ECMO was weaned rapidly post coiling with decannulation in under 24 hours. The patient is currently stable in medically managed compensated heart failure 4 years postoperatively.

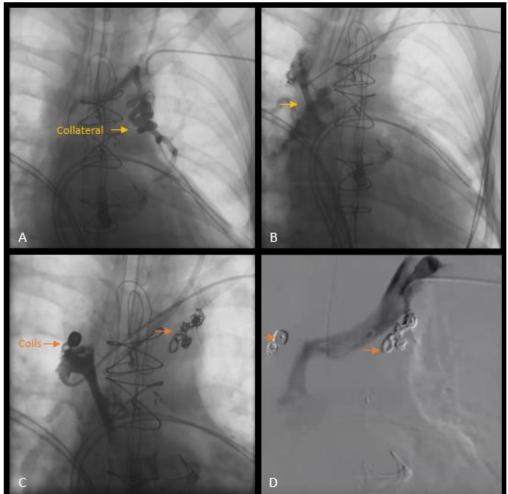


Figure 1: Right heart catheter demonstrating significant veno-venous connections from the innominate vein to pulmonary veins. A) Collateral to the left pulmonary vein. B) Collateral to right pulmonary vein. C) Multiple Boston Scientific Interlock[™] coils. D) Patent Innominate vein post-coiling with no residual fistulous venous connections.

Discussion:

Advances in the diagnosis, medical management and surgical interventions for congenital heart disease has improved survival into adulthood for many patients (4). Adult congenital heart disease (ACHD) patients have unique anatomy and complex physiology that is not cured by current interventions. These patients require highly specialized medical and surgical services with experience in ACHD, including adult congenital cardiology, interventional cardiology, cardiac anesthesiology, intensive care, perfusionist, ECMO services, and congenital cardiac surgeons. This breadth of knowledge, with multidisciplinary teamwork was critical for the successful management of our patient.

Veno-venous collaterals are a known phenomenon in patients with a Fontan circulation (1). The benefits of coiling chronically present venovenous collaterals are debated, as not all venous collaterals cause shunting and result in hypoxia. The indication for collateral coiling is often if they are large and cause resting hypoxia (3). However, when to intervene and how many collaterals to coil is unclear. This patient had no evidence of collaterals on preoperative investigation or visible intraoperatively. We believe the patient had collaterals present that were dormant preoperatively. They opened creating significant shunting due to an acute change in pathophysiology postoperatively. Significant opening of collateral vessels postoperatively caused refractory hypoxia in our patient.

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These were rapidly identified and coiled, leading to successful VV-ECMO weaning with correction of hypoxia.

Conclusions:

This case demonstrates successful weaning from VV-ECMO using embolisation for veno-venous collaterals with significant right to left shunt and hypoxia, in a patient with Fontan circulation. Acute refractory decompensation rapidly resolved following embolisation. Venous collateral coiling should be considered in the context of clinical condition and hypoxia. A low threshold is required to investigate for collateral vessels even when they are not present in the preoperative phase.

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Conflict of interest:

none declared.

Author contributions:

Charlotte C. Frost: Manuscript writing & editing.

Vinod Sharma: Manuscript editing.

Dale Murdoch: Manuscript editing.

Douglas Wall: Manuscript editing.

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