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Transcatheter creation of a pulmonary artery to left atrial fenestration in a failing Fontan circulation using the Atrial Flow Regulator (AFR)

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Brief Report

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Abstract

Transcatheter creation of an interatrial communication using the Occlutech Atrial Flow Regulator Device for pulmonary hypertension or heart failure is well described. We report a case of an 8-year-old boy with a failing Fontan circulation, in whom the Atrial Flow Regulator was used to successfully create a fenestration between the pulmonary artery and left atrium, improving his clinical condition.

Many complex cyanotic congenital heart defects may be palliated in a staged manner, leading to a Fontan procedure. Although modifications of this operation have improved short and medium-term survival, high impedance from pulmonary vascular resistance results in systemic venous congestion, low cardiac output, and failure of the Fontan circulation.[1] Surgical fenestration limits systemic venous pressure, decreases systemic venous congestion, and improves cardiac output.[1] Complications of a failing Fontan circulation, including plastic bronchitis and protein-losing enteropathy, may be treated by transcatheter recanalisation or stenting of an occluded surgical fenestration.[2]

We describe the first paediatric case, in whom the Occlutech Atrial Flow Regulator device (Fig 1; Occlutech International, Helsingborg, Sweden) was used to create a fenestration in a Fontan circulation by direct communication between the pulmonary arteries and left atrium in a patient with protein-losing enteropathy.

Case report

An 8-year-old boy with double inlet left ventricle, ventricular septal defect, and interrupted aortic arch underwent staged single-ventricle palliation (Norwood-type procedure with aortic arch reconstruction and creation of a left-sided modified Blalock–Taussig shunt, Hemi-Fontan procedure), culminating in a Fontan operation using a 4 mm fenestrated lateral tunnel at 3 years old. He remained well for 4 years, but became increasingly lethargic with generalised oedema, hepatomegaly, ascites, and poor weight gain. Resting oxygen saturation was 91%, likely due to small systemic-to-pulmonary venous collateral vessels. The fenestration was noted to be patent. The albumin level was 21 g/L (range 40–52 g/L) with a faecal alpha-1 antitrypsin level of 5.09 mg/g (upper limit of normal 0.48 mg/g), confirming protein-losing enteropathy. He was treated with Furosemide and Lisinopril. One year later, echocardiography and MRI catheter demonstrated an occluded fenestration, and proximal left pulmonary artery narrowing. Central venous pressure was 12 mmHg. The pulmonary vascular resistance at baseline was elevated at 6.7 WU.m^2 decreasing to 2.7 WU.m^2 with inhaled nitric oxide and 100% oxygen. He commenced Sildenafil, Octreotide, and Budesonide, but was readmitted 1 week later with severe leg and scrotal oedema.

Recanalisation of the previous fenestration was attempted from the femoral venous approach. It was not possible to cross the surgical fenestration, so puncture of the GoreTex baffle using a trans-septal needle under transoesophageal echocardiographic guidance was performed and a 6 mm × 18 mm Valeo stent (Bard Inc., New Province, NJ, United States of America) was implanted without pre-dilatation of the baffle puncture. The left pulmonary artery was stented with a 10 mm × 26 mm Valeo stent. Pre-procedure distal mean left pulmonary artery and mean right pulmonary artery pressures were 13 mmHg and 19 mmHg, respectively. Post-procedure, the distal mean left pulmonary artery pressure and lateral tunnel pressure were 18 mmHg, left ventricular end-diastolic pressure 8 mmHg, and left atrial pressure 9 mmHg. Oxygen saturation decreased from 95 to 86%.

The following day oxygen saturation increased to 96% with no flow across the stent on transoesophageal echocardiography and saline bubble contrast, presumed to be due to stent thrombosis. He was treated with Alteplase and heparin without improvement after 48 hours.

Figure 1. Occlutech Atrial Flow Regulator device.

Twenty-four hours after stopping Alteplase, repeat cardiac catheterisation was performed to create a new fenestration because of a possible high risk of systemic embolisation of thrombus associated with attempted recanalisation of the Valeo stent. Because of the possible increased risk of bleeding, surgical support and cardiopulmonary bypass facilities were on standby. Using right femoral and right internal jugular access, angiography demonstrated a patent left pulmonary artery stent, but complete occlusion of the stent between the lateral tunnel and left atrium (Fig 2a). Due to the close proximity of the pulmonary artery and the left atrium, a BRKTM needle (Abbott Medical, Chicago, United States of America) was passed through a 7 Fr Performer™ Guiding Sheath (Cook Medical LLC, Bloomington, United States of America) from the right internal jugular vein through the pulmonary artery into the left atrium under transoesophageal echocardiographic and fluoroscopic guidance. It was thought that previous multiple sternotomies resulting in adhesions would reduce the risk of bleeding into the pericardium. Once the left atrial pressure tracing was recorded, the Performer™ sheath was advanced into the atrium. A 5 Fr Judkins right coronary catheter and 0.035″ hydrophilic wire crossed from the atrium through the left ventricle to the descending aorta and was exchanged for a 0.035″ Amplatz Extra-Stiff Wire (Cook Medical LLC). The newly created communication was pre-dilated with a 10 mm × 2 cm Conquest Balloon (Bard Inc.), followed by a 12 mm × 2 cm Atlas Gold Balloon (Bard Inc.) (Fig 2b). The jugular venous sheath was upsized to 16 Fr and advanced towards the floor of the atrium. A 10 mm, medium height Occlutech Atrial Flow Regulator™ was deployed under echocardiographic and fluoroscopic guidance (Fig 2c and d). Oxygen saturation immediately reduced from 95 to 73%. Once satisfactory position and flow were confirmed, the Occlutech Atrial Flow Regulator was released. The mean gradient measured on transoesophageal echocardiography through the device was 4 mmHg (Fig 2e). He was extubated the same evening and started on an intravenous heparin infusion, followed by a combination of Aspirin and Warfarin. Complete atrioventricular dissociation is seen at the end of the procedure resolved after 6 days of steroids. Oxygen saturation remained between 70 and 80%. Two weeks later, the ascites and scrotal oedema had resolved, weight decreased from 20.5 kg to 15.9 kg and albumin increased from 21 g/L to 44 g/L (range 40–52 g/L).

The patient was discharged home after 3 weeks. At 3-month follow-up, echocardiography demonstrated good flow through the Atrial Flow Regulator device with a maximum velocity of 1 m/s and a mean gradient of 2 mmHg. The resting saturation was 71% without oxygen supplementation. Haemoglobin concentration was elevated at 220 g/L.

Discussion

Reported survival of patients with Fontan circulation at 20 years is 70–85%.[3] Eventually, all patients will develop failure of the Fontan circulation.[3] Treatment options include cardiac transplantation or palliation by decompression of the Fontan pathway. Protein-losing enteropathy occurs in 10–20% and plastic bronchitis in 1–14% of patients, 1 month–20 years later.[4] Five-year survival of these is 46% and 40–78% respectively.[4]

The optimal management of the failing Fontan circulation remains controversial. A patent fenestration between the systemic venous return and atrial mass may reduce systemic venous congestion and improve cardiac output at the expense of oxygen desaturation.[5] An occluded surgical fenestration can be recanalised and stented or a new fenestration created using a baffle puncture and stent placement.[6] Thrombotic occlusion and/or systemic embolisation are frequent complications, particularly if the fenestration diameter is less than 5 mm.[7]

The Atrial Flow Regulator device is designed for patients with advanced heart failure or pulmonary hypertension to create an interatrial communication and decompress either the left or right atrium.[8] To our knowledge, this is the first case where the device has been used to create a fenestration between the pulmonary artery and left atrium in a paediatric patient with failing Fontan circulation. It has been used in an adult patient with failing Fontan circulation and in four children to decrease the fenestration size.[5,9,10] This is an off-label use and procedural success without complication may be unpredictable if there is no surgical scarring or adhesions in the space between the pulmonary artery and atrium. The Atrial Flow Regulator has a larger ratio of fenestration to disc diameter than fenestrated atrial septal defect devices and no thrombogenic patch material.[5] This may reduce reocclusion rates. It ensures a predictable fenestration with controlled blood flow to decompress the central venous system. In this case, it provided a better clinical outcome and a reliable patency of the fenestration than the initial stenting procedure.

Although a 10 mm AFR might be considered large for a patient with a Fontan circulation, this size was chosen to provide reliable Fontan circuit decompression. We were also prepared to downsize this fenestration by implanting a smaller AFR within this if the

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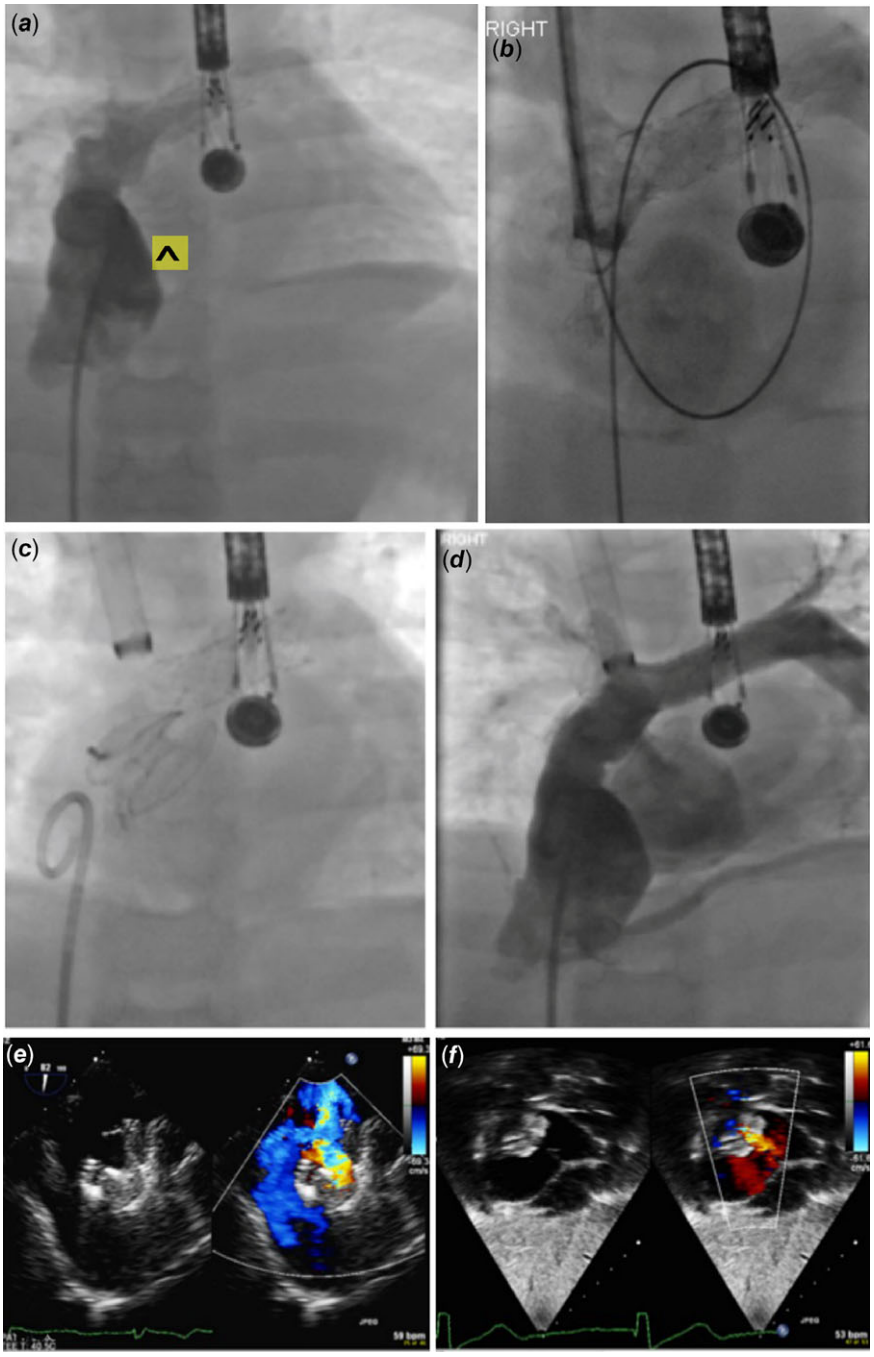

Figure 2. (a) Angiogram performed via femoral venous access with catheter placed in lateral tunnel showing no flow through the baffle stent with contrast injection (**indicated by ^**); (b) Angiogram after Atlas balloon dilation of the neofenestration demonstrating no extravasation of contrast; (c) and (d) Angiogram performed via femoral venous access with catheter in lateral tunnel showing Atrial Flow Regulator device and flow through it with contrast injection; (e) Transoesophageal echocardiogram during the procedure showing Atrial Flow Regulator device with good flow through the device; (f) Transthoracic echocardiogram the day after the procedure showing Atrial Flow Regulator device with good flow through the device.

patient had not tolerated desaturations. If a smaller AFR had been implanted initially, and the fenestration needed to be enlarged, or further decompression, technically this would have been more difficult requiring stenting. The 16 F sheath also allowed for possible easier retrieval and repositioning of the device if that had been required.

Conclusion

Failure of the Fontan circulation during long-term follow-up is common. A surgical fenestration reduces systemic venous congestion and subsequently increases cardiac output, but may occlude spontaneously. This case used an Atrial Flow Regulator device to create a new fenestration and treat elevated systemic venous pressures. This novel technique may be used increasingly for future palliation of these complex patients.

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Conflicts of interest. Shakeel Qureshi is a consultant for Occlutech.

Ethical standards. The Institutional Review Board deemed that the anonymised retrospective case review to be exempt from requiring formal approval.

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