

Fontan circuit fenestration occlusion with Occlutech patent ductus arteriosus occluder

Zongezile Masonwabe Makrexeni and Lungile Pepeta

Division of Paediatric Cardiology, Paediatrics and Child Health, Dora Nginza Hospital, Walter Sisulu University, Port Elizabeth, South Africa

Address for correspondence:

Dr Z.M. Makrexeni
Division of Paediatric Cardiology
Paediatrics and Child Health
Dora Nginza Hospital
Walter Sisulu University
Port Elizabeth
6001
South Africa

Email:

mmmaso30@gmail.com

INTRODUCTION

William-Beuren syndrome is due to a microdeletion in the chromosome region 7q11.23, and commonly presents with progressive supra-aortic stenosis and non-progressive or improving pulmonary artery stenosis.⁽¹⁾ It is unusual for this syndrome to present with a single ventricle (univentricular) congenital cardiac aberration. Management of a univentricular heart traditionally involves staged shunting – including a Blalock-Taussig Shunt, a Glenn Shunt (with a Blalock-Taussig Shunt take-down), and completion of a total cavo-pulmonary connection (TCPC, “Fontan circuit”) with or without a right atrium-to-TCPC fenestration. The introduction of fenestration at the time of completion of the “Fontan circuit”, was a significant advance in the peri-operative care of patients with univentricular physiology. In the immediate post-operative period, the ability to shunt blood from right to left through the fenestration reduces volume and pressure load on the pulmonary circulation and augments systemic cardiac output – thereby reducing morbidity and mortality, while awaiting the cardiovascular system’s adaptation to the new haemodynamic situation.⁽²⁻⁴⁾ However, in the long-term, the fenestration is a source of continuing systemic desaturation, and a potential conduit for paradoxical thrombo-embolisation.^(5,6) Device closure of a Fontan fenestration is a widely accepted strategy to eliminate these potential complications.⁽⁷⁾ It is safe and associated with

ABSTRACT

We present a case of Fontan fenestration closure using an Occlutech patent ductus arteriosus occluder. The patient was diagnosed with tricuspid atresia, pulmonary atresia and secundum ASD at birth. He had a bilateral modified Blalock-Taussig shunt to promote growth of hypoplastic branch pulmonary arteries. He had a bidirectional Glenn Shunt at 11 years of age, and a total cavo-pulmonary connection (TCPC) with a fenestration at 19 years of age. He presented a year later with severe exercise intolerance and cyanosis. Cardiac catheterisation showed hypoxemia, normal pulmonary pressures, patent Fontan circuit and branch pulmonary arteries, and a Fontan circuit-to-right atrium fenestration with right to left shunting. A short shank Occlutech PDA occluder with a waist of 8mm, a pulmonary end of 10mm, an aortic disk of 16mm, and a length of 7mm, was selected to close the fenestration through the transvenous approach. The fenestration achieved complete immediate occlusion with this device. SAHeart 2018;15:210-212

acute and persistent improvements in oxygen saturation, somatic growth, exercise capacity, and reduction in anticoagulant requirements.⁽⁸⁻¹⁰⁾ There are no commercially available devices that are designed specifically for Fontan fenestration occlusion, except for the PFO Star device described by Boshoff, et al.⁽⁵⁾ The most commonly used device is the Amplatzer® Septal Occluder (ASO; St Jude Medical, St Marks, MN).^(7,11,12) We report a case of successful closure of Fontan fenestration using an Occlutech patent ductus arteriosus (PDA) closure device (Occlutech®, Helsingborg, Sweden).

CASE PRESENTATION

We present a 21-year-old male patient, who presented with dysmorphism and severe cyanosis at birth. He was diagnosed with William-Beuren syndrome, congenital tricuspid atresia, and pulmonary atresia with a secundum atrial septal defect (ASD). The patient had bilateral modified Blalock-Taussig shunts, as he had hypoplastic branch pulmonary arteries. He had a bidirectional Glenn Shunt at 11 years of age, and a TCPC with a Fontan circuit-to-right atrium fenestration done when aged 19 years. A year later, the patient presented with easy fatigability, exercise intolerance, and severe cyanosis with

oxygen saturation of 84% in room air. The echocardiogram showed a patent TCPC with a fenestration shunting right to left. There was also a high haemoglobin concentration of 16.7g/dl, and low arterial blood gas oxygen saturation of 89%.

Under general anaesthesia, both venous and arterial access were achieved. Hypoxemia was confirmed in that the systemic arterial saturations remained at 89%, with systemic venous saturations very low at 54%. The pulmonary artery pressures were normal, with a mean pressure of 13mmHg, and there was no indication of obstruction in the branch pulmonary arteries. The inferior vena cava venogram confirmed that the TCPC was patent with a fenestration that was shunting from right to left, and measuring 4.6mm in diameter. The superior vena cava venogram confirmed a patent Glenn Shunt and branch pulmonary arteries (Figure 1A). Based on the size of the defect and as per the manufacturer's guidelines (Occlutech®, Helsingborg, Sweden), an Occlutech® PDA occluder with a waist of 8mm and pulmonary end of 10mm, was selected to close the fenestration (Figure 1B). This is a short shank (with a short length) device that will endothelialise in 6 months, and will not cause turbulence in this low-pressure system. Another reason why this device was chosen, is that it is much cheaper compared to the commonly used ASO (Occlutech®, Helsingborg, Sweden; St Jude Medical, St Marks, MN). We used the device itself for test occlusion, instead of a balloon interrogation. We did an angiogram before releasing the device from the delivery wire. When we were satisfied that the device had closed the defect completely, the device was released. The fenestration was successfully closed, with no residual shunting across it (Figure 1C). Following fenestration closure, the sys-

temic arterial saturations improved immediately from 89% - 100% in room air. The patient showed clinical improvement following fenestration closure, in that he was asymptomatic and remained with oxygen saturation of more than 95% in room air on follow-up.

DISCUSSION

Device closure of a Fontan fenestration is a widely adopted procedure. It is safe and associated with immediate and sustained overall clinical improvement and reduced needs for anticoagulation.⁽⁸⁻¹⁰⁾ The Amplatzer® septal occluder has been used for a long time to occlude Fontan fenestrations – most probably due to its longstanding availability and because the fenestration might be considered to be a neo-atrial septal defect.^(7,11) The Amplatzer® duct occluder type 2 has also been used with success.^(11,12) The device seems well suited for Fontan fenestration closure, as it is a soft, flexible device that can conform to a variety of fenestration lengths and angles, and the discs are known to flatten with time, enabling them to follow the contour of the GORE-TEX conduit in the total cavo-pulmonary circulation. It is also a low profile device, with most device sizes deliverable through concomitant low-profile delivery sheaths (4Fr and 5Fr delivery systems).⁽¹²⁾ The new Occlutech® paravalvular leak device (Occlutech®, Helsingborg, Sweden) has been used for the closure of Fontan fenestration.⁽¹³⁾ This device offers a combination of a small delivery sheath size, high flexibility, less material, and a low profile.

The Occlutech® duct occluder (Occlutech®, Helsingborg, Sweden) has a meshwork of self-expandable nickel and titanium (nitinol) alloy wire. It has a “champagne cork” configuration.

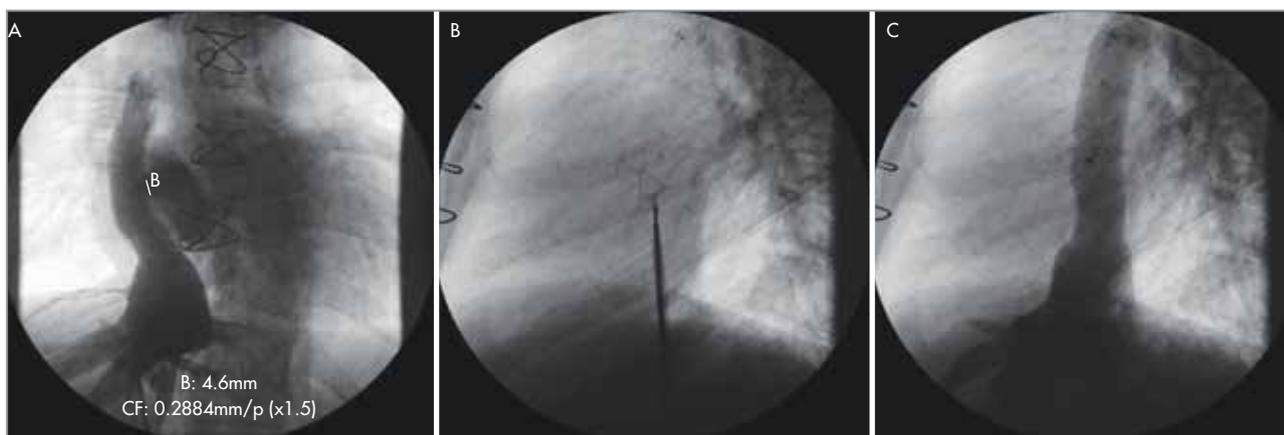


FIGURE 1: Leaflet involvement versus successful repair.

- A. Fontan circuit conduit angiogram showing patent conduit and conduit-to-right atrium fenestration measuring 4.6mm.
- B. Occlutech duct occluder deployment in the fenestration.
- C. Complete fenestration occlusion with Occlutech duct occluder in situ.

There is a flat disk on the aortic side, which is continuous with the body ("shank") of the device through nitinol braiding. The aortic side of the shank measures 3.5mm - 14mm in diameter and "flares up" at the pulmonic end – thus giving it a diameter 1.5mm - 4mm larger than the aortic end of the shank (Figure 1B). The device length varies from 4.25mm - 16mm. Embedded in the body are polyethylene terephthalate (PET) threads that are expected to enhance defect closure rate. These devices are delivered through 6F to 9F Cook's delivery system, depending on device size. This is the first case report, to the best of our knowledge, of the use of a Occlutech® PDA occluder for Fontan circuit fenestration occlusion.

CONCLUSION

A Fontan circuit-to-right atrium fenestration has become standard peri-operative management of single-ventricle heart conditions undergoing total cavo-pulmonary circulation palliation. Percutaneous closure of the fenestration with various devices has also become standard management. Percutaneous closure of this defect with an Occlutech® PDA occluder, expands the armamentarium of devices that can be used for closure of this defect. The device is much more cost-effective compared to other devices used for this purpose.

Conflict of interest: none declared.

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