Cardiology in the Young (2013), **23**, 766–768
doi:10.1017/S1047951112001886

Brief Report

Stent therapy for acute and chronic obstructions in extracardiac Fontan conduits

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Abstract We describe transcatheter therapy for early onset occlusion or stenoses of extracardiac conduits in three children who had undergone Fontan completion. Successful stent implantation was associated with complete resolution of symptoms.

Keywords: Fontan; extracardiac conduit; stents

Received: 13 June 2012; Accepted: 16 September 2012; First published online: 20 November 2012

Since its initial description, the extracardiac conduit has been extensively used for completion of the Fontan operation in children with univentricular hearts.[1] It has the advantage of avoiding surgical suture lines in the atrium and therefore potentially reducing future arrhythmia risk, of optimised fluid dynamics, and the operation is technically easy to perform.[2–4] Follow-up studies have demonstrated a mild reduction in the internal diameter of the conduit within the first few months after surgery, with no significant late stenosis at up to 15 years after Fontan completion.[5,6] We describe the cases of three young patients with evidence of acute or chronic conduit obstruction, which was successfully relieved by percutaneous stent implantation.

Patient 1 was a 5-year-old girl who presented with acute abdominal pain 3 months after Fontan completion for tricuspid atresia, using a 16-mm diameter Gore-Tex conduit (Distributor W.L. Gore & Associates BV, Tilburg, The Netherlands). Angiography demonstrated thrombotic occlusion of the conduit (Fig 1). The occluded segment could be crossed with a 0.035-inch guidewire and catheter combination via a femoral venous access. Recanalisation of the conduit was successfully achieved by implantation of a Palmaz stent (Cordis Corporation, Miami Lakes, Florida, United States of America), which was dilated up to 15 mm.[7] Following subsequent thrombolytic therapy, the patient recovered uneventfully. There was no evidence for a coagulopathy. Over a follow-up of over 5 years, she has had no further complaints.

Patient 2 was a 3-year-old girl who developed progressively worsening cyanosis within 3 months after Fontan completion, using an 18-mm conduit, for visceral heterotaxy syndrome and complex univentricular heart. She had had epicardial pacemaker implantation during Fontan completion, for treatment of sinus node disease. Angiography via the femoral and jugular veins demonstrated a localised stenosis at the junction between the conduit and inferior caval vein. This was successfully treated with implantation of a Palmaz stent, which was dilated to 18-mm in diameter using a high-pressure balloon (Atlas, Bard Peripheral Vascular Incorporation, United States of America) dilated up to 15 atmospheres (Fig 2). Multiple collateral vessels to the pulmonary venous atrium were also occluded successfully, resulting in an improvement in the transcutaneous oxygen saturation from 78% to 94% at discharge from hospital (Fig 2).

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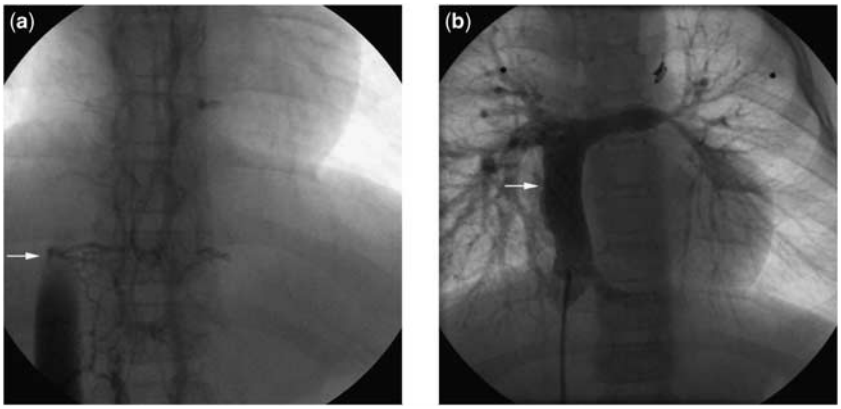

Figure 1.
*(**a**) Hand injection of contrast in the femoral vein shows complete occlusion of the extracardiac conduit (arrow, reprinted with permission). (**b**) Contrast injection following stent implantation (arrow), confirming free flow through the conduit (reprinted with permission).*

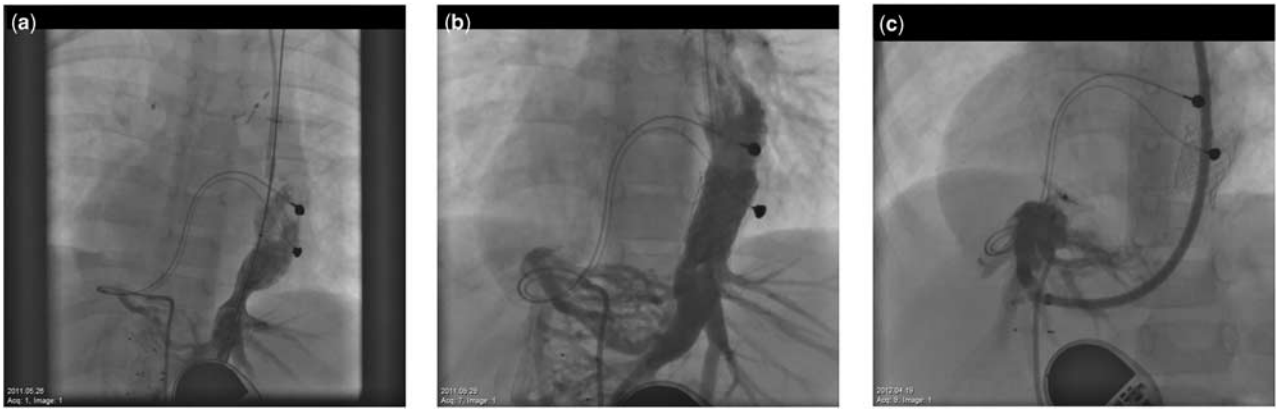

Figure 2.
*(**a**) Contrast injection in the extracardiac conduit confirming a stenosis at the junction with the inferior caval vein in patient 2. (**b**). The result after stent implantation in patient 2. (**c**) The extensive collateral network to the pulmonary venous atrium in patient 2 has been occluded with a 10 mm Amplatzer muscular ventricular septal defect occluder.*

Patient 3 was a 2.5-year-old boy with complex univentricular heart, who presented with protein-losing enteropathy, confirmed by stool alpha-1 antitrypsin clearance studies, 5 months after Fontan completion with an 18-mm conduit. Angiography via the femoral vein demonstrated a mid-conduit stenosis, without any measurable pressure gradient (Fig 3). This was treated by stent implantation (Palmaz), with dilation of the stent to 18 mm in diameter using a high-pressure balloon (Atlas). The procedure was uneventful. Within 6 weeks after the procedure, the patient had normal serum albumin levels and normal stool alpha-1 antitrypsin levels.

Comment

The extracardiac Fontan operation is associated with excellent long-term survival, and a small reported incidence of conduit-related problems.[2–6] Optimal size of the conduit and the optimal age for Fontan completion are still debated, as it is likely that patients who have not realised their growth potential may require conduit replacement in later life owing to the absence of growth in the diameter of the conduit. The conduit sizes in the patients described here – 16 mm versus 18 mm – were chosen by the surgeon on the basis of the diameter of the inferior caval vein at the lower anastomotic site. There are few reports of stent therapy for conduit failure, and protein-losing enteropathy may not be reversible in some patients despite successful therapy of conduit stenosis.[8,9] Early onset of new symptoms should, however, prompt aggressive investigation for potentially treatable causes of failure of the Fontan circulation. Our current institutional policy is to maintain anticoagulation using coumadin in all patients who have undergone Fontan completion – with or without stent implantation in the

https://doi.org/10.1017/S1047951112001886 Published online by Cambridge University Press

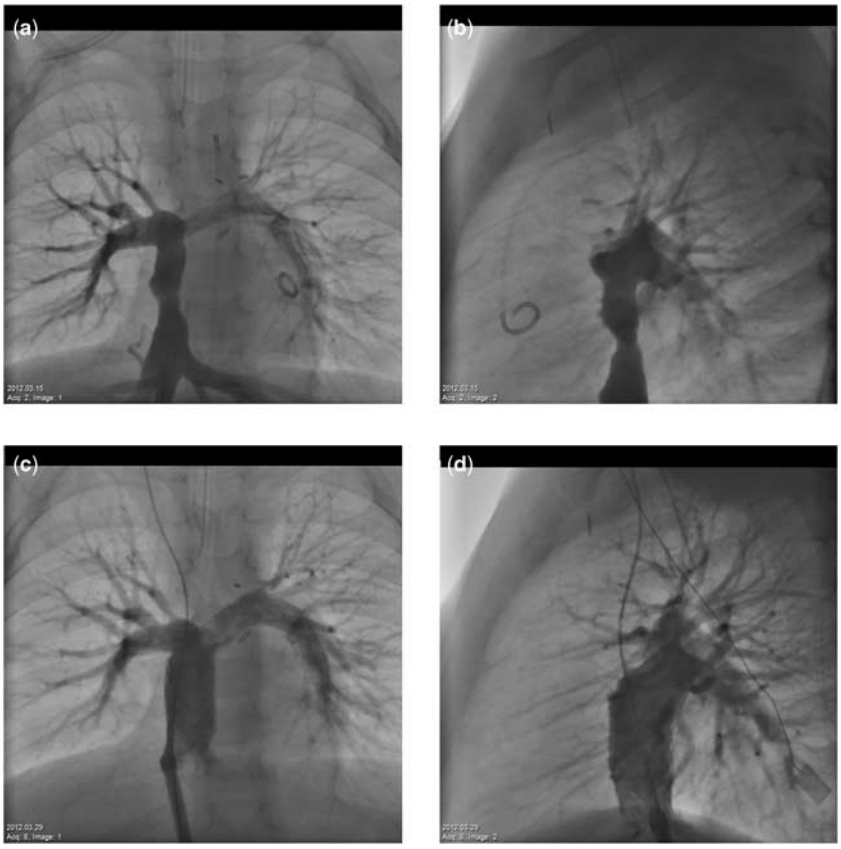

Figure 3.
*(**a** and **b**) Antero-posterior and lateral angiographic projections in patient 3, demonstrating mid-conduit stenosis. (**c** and **d**) Antero-posterior and lateral projections following stent implantation in patient 3.*

Fontan circuit – to maintain the international normalised ratio at between 2.5 and 3.0.

Acknowledgements

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