Percutaneous Stenting of Dacron Conduit Stenosis for Extracardiac Fontan Procedure

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Abstract

We describe a case of successful transcatheter therapy with covered stents for chronic stenosis of extracardiac conduit in a patient who had undergone Fontan completion. Successful stent implantation was associated with complete resolution of symptoms.

Keywords: Percutaneous Stenting; Dacron Conduit Stenosis; Fontan Procedure

Introduction

The extracardiac conduit is actually considered the treatment of choice for children with univentricular hearts. However, The development of stenoses later in the postoperative period is possible due to narrowing in the areas of anastomosis. We describe the case of a young patient with evidence of chronic conduit obstruction, which was successfully relieved by percutaneous stent implantation.

Case Report

A female patient with tricuspid valve atresia, malposition of great vessels and pulmonary pathway stenosis (Type II b) underwent a Blalock anastomosis at the age of 2 weeks. In 2004 (10 years old) she underwent a single stage extra-cardiac total cavopulmonary connection using a 21 mm Dacron tube. She had initial excellent outcome with good hemodynamics and a SPO2 of 96%.

Six years after she presented with ascites and hepatomegaly due to thrombosis of the inferior vena cava. Anamnesis revealed a non-observance for the anticoagulant therapy. She was successfully managed with heparin and then acenocoumarol. Than the girl was lost sight of for 7 years.

In August 2017, she presented with anasarca, enormous ascites (Figure 1) due to Protein-Losing-Enteropathy (PLE) as evidenced by the lab results: albumin 12 g/l; total protein 26 g/l, her alpha-1 antitrypsin stool concentration was significantly elevated (13 mg/g; normal 0 - 2 mg/g).

Figure 1: Enormous ascites because of failing Fontan.

The patient said that she was taking her anticoagulant treatment correctly during the last period. Her International Normalized Ratio (INR) was 2.6.

Transthoracic echocardiography revealed good ventricular systolic function, a moderate systemic leaflet insufficiency, a permeable Glenn. The Extra cardiac conduit (ECC) wasn’t well visualized.

Cardiac catheterization was performed to investigate the morphology of the ECC patency. It was done under general anaesthesia, after informed consent was obtained.

Angiography via the femoral vein using the anteroposterior and lateral projections demonstrated a localised stenosis of more than 45% at the junction between the conduit and inferior caval vein and a second stenosis in the medium part of the ECC (Figure 2). A minimal pressure gradient (2 mm Hg) across the ECC was found. This was successfully treated with implantation of one first stent (Andra XXL 43 mm) at the distal lesion (VCI-ECC) and a second stent was implanted during the same procedure, Andra stent XL 30 mm. Both stents were dilated to 21 mm dilated to 18-mm using a high-pressure balloon (Figure 3). The conduit diameter increased significantly after stent placement. The procedure was uneventful. Oral anticoagulation was stopped 1 week before, and restarted 1 day after stenting. Intravenous heparin was given during (100 units/kg) and for 24 hours following (200 units/kg/24 hours) the procedure.

Within 8 weeks after the procedure, the patient had normal serum albumin levels and normal stool alpha-1 antitrypsin levels. Ascites disappeared significantly (Figure 4).
The extra cardiac conduit (ECC) is currently the preferred surgical strategy to palliate univentricular hearts. Compared to the classic Fontan operation it has the advantage of reducing future arrhythmia risk [1]. However, stenosis especially at the sites of anastomoses may occur leading to under optimal hemodynamic conditions and to a “Failing Fontan” situation. That has increasingly been reported in some cases. However the incidence seems to be very rare in some retrospective studies of early and mid-term follow up [2].

The mechanisms of ECC stenoses are likely multifactorial. It is frequently observed in patients with small conduits of 16 - 18 mm because of lack of growth of the prosthetic material. The case we are presenting is extremely rare because of the relative huge conduit (21 mm). The thrombogenicity and biochemical aspects of the graft material Dacron may be an explication. In fact, Excessive neointima proliferation of the entire graft lumen has previously been reported in stenosed ECC Dacron grafts [3]. Dacron grafts are described to be at high risk of stenosis both early and late after surgery when compared to Gore-Tex conduits. Udink ten Cate FEA., et al. [4] found a strong correlation between right pulmonary artery size and stenosis diameter of patients with a mid-distal pathway obstruction [4].

Diagnosis of the stenosis is not evident in the low-pressure cavopulmonary system, where lesions may be clinically relevant even in the absence of a measurable pressure at catheterization. That was the case of our patient. Historically Most Fontan patients with a significant ECC obstruction undergo redo surgery. Transcatheter treatment of the ECC narrowing may improve the hemodynamics in Fontan circulation, even in patients without measurable pressure gradient [5]. Furthermore, Cate FEA., et al. [4] in their retrospective study about 19 cases, reported that it was possible to treat a variety of forms of obstruction, with few adverse events and a good outcome.

Conclusion
A significant pathway stenosis should be excluded in all Fontan patients presenting with PLE or failing Fontan haemodynamics. Percutaneous stent implantation is feasible and safe and may provide a good alternative to early reoperation.

Conflicts of Interest
The authors declare that there are no conflicts of interest.
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Bibliography


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