Stent Implantation in Surgical Cavopulmonary Conduit: Report of Two Cases

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Despite advances in Fontan surgery, obstruction in extracardiac ducts may occur and cause clinical deterioration. We report two cases of stent implantation for correction of stenosis in the Fontan surgery. Ascites was a common clinical sign; a patient was protein losing enteropathy. All procedures were angiographically and clinically successful.

Introduction

The Fontan procedure allowed the physiological correction with the separation of pulmonary and systemic circulations in children with congenital heart disease, in which biventricular repair is not possible. The criteria for the indication of Fontan surgery are well established in literature[1]. Blood flow free of obstructions in the pulmonary arteries and systemic veins, as well as low pressure in the pulmonary arteries are important factors for maintaining optimal hemodynamic conditions in the long term[2]. However, the development of stenosis in the clinical follow-up, especially at the sites of anastomoses may occur, leading to underoptimal hemodynamic conditions[3]. The most common clinical manifestations observed in these cases are: ascites, pleural effusion, protein-losing enteropathy (PLE)[4].

This article aims to illustrate postoperative complications that can be circumvented in a minimally invasive way, assisting in the management of children with complex congenital heart disease undergoing univentricular correction[5].

Case report

Case 1

Patient GTM, ten years and six months old, with tricuspid atresia, in late postoperative of Fontan surgery with extracardiac conduit (ECC) Gore-Tex (GT; W.L. Gore & Associates, Flagstaff, AZ, USA) held on 07.20.2003 when patient was six years and nine months. Admitted on April 25, 2007 with a picture of PLE, the hemodynamic study revealed moderate stenosis in the anastomosis of the inferior vena cava (IVC) with the Fontan tube (measuring 9 mm in stenosis and and 18 mm as a reference diameter), with a gradient in 2 mmHg at rest (Figure 1). The patient underwent implantation of Cheatham Platinum stent 8 ZIG 39 mm (NuMed, Inc., Nicholville, NY, USA) with BIB balloon 14x45 mm (NuMed, Inc., Nicholville, NY, USA), and post-balloon dilatation Max LD (Cordis Corp., Miami Lakes, Florida, USA) of 18x40 mm at the site of stenosis, successfully, without residual gradient. He was discharged on the 11th post-implantation with remarkable improvement of clinical condition. The treatment resulted in stability of albumin levels and allowed the child's return to school activities.

Six months of clinical stability evolved, while the symptoms of PLE have gradually returned. After 11 months, another hemodynamic study revealed good patency of the stent and no signs of thrombosis or restenosis. The pulmonary artery mean pressure (PA) was 16 mmHg.

On June 02, 2008, with favorable conditions for surgery, the patient underwent surgery for the exchange of ECC, now with fenestration (due to maintenance of high PA). Patient was discharged on 21st postoperative day for clinical follow-up, remaining without readmission.

Case 2

Patient J.A.P., six years old, with cyanotic congenital heart disease: Left Ventricle Type Single Ventricle Double Inlet Way and surgical history of Blalock-Taussig anastomosis on the fourth day of life, Glenn two years old and Fontan surgery five years old (bovine jugular vein ECC). She was admitted on 04/04/2007 with chronic cough and hemoptysis in the last 24 hours. Hemodynamic study with pulmonary arteriography revealed narrowing of the right IVC-PA tube without pressure gradient and a large thrombus in the pulmonary branches to the base of the left lung (pulmonary embolism). Under oral anticoagulation, there was significant improvement of clinical symptoms, and the patient was discharged after 12 days without an image suggestive of thrombus in ECC on echocardiogram.

On August 28, 2008, she was hospitalized due to a condition of ascites; abdominal ultrasonography revealed dilatation of suprahepatic veins and mild splenomegaly. She underwent catheterization in which were found diffusely stenotic ECC (smaller diameter = 4 mm and reference diameter = 14 mm - Figure 2A) with PA-IVC gradient of

Keywords

Stents; ascites; protein-losing enteropathies; heart defects, congenital.

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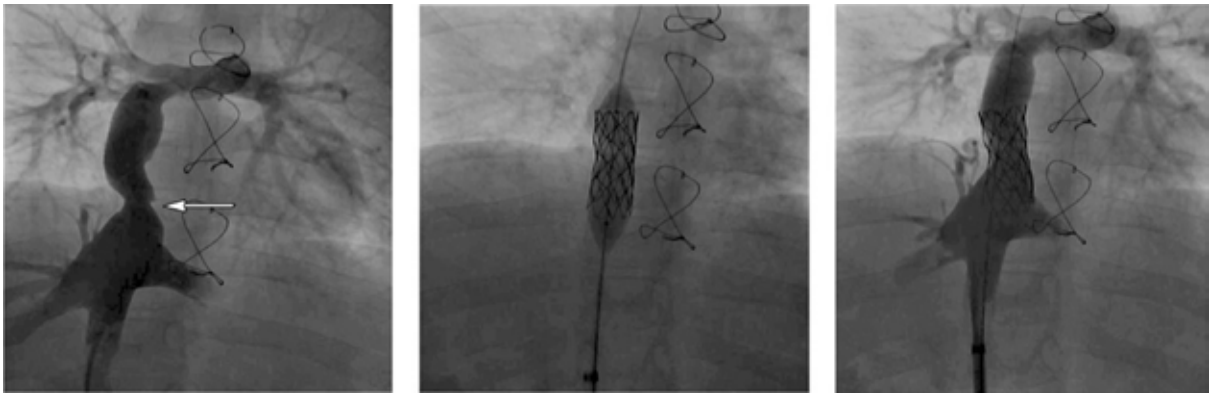

Figure 1 - Angiographic aspects pre: IVC-ECC stenosis (arrow) and post: stent implantation.

8mmHg and pulmonary artery mean pressure of 15 mmHg. Palmaz-Schatz stent placement (Cordis Corp., Miami Lakes, Florida, USA) 4014 in LD Max balloon (Cordis Corp., Miami Lakes, Florida, USA) of 14x40 mm, with partial success due to a residual gradient of 6 mmHg. In three days, the patient was referred to stent redilation with two simultaneous Power Flex balloons (Cordis Corp., Miami Lakes, Florida, USA) of 9x20 mm and 10x20 mm - 14 atm (Figure 2B), with disappearance of the gradient. The hospital development was favorable with resolution of symptoms under oral anticoagulation and sildenafil. The patient is well on follow-up.

Discussion

The long-term stability of Fontan surgery depends on the satisfactory development of the pulmonary circulation during the somatic growth of children operated early and maintaining a good ventricular function. The development of stenoses in the postoperative period is possible due to narrowing or tensions in the areas of anastomosis or extrinsic aortic compression, as observed by some authors[6].

We report two cases where univentricular corrections were made with different techniques, and changes in flow caused by stenosis at the level of the conduits were important

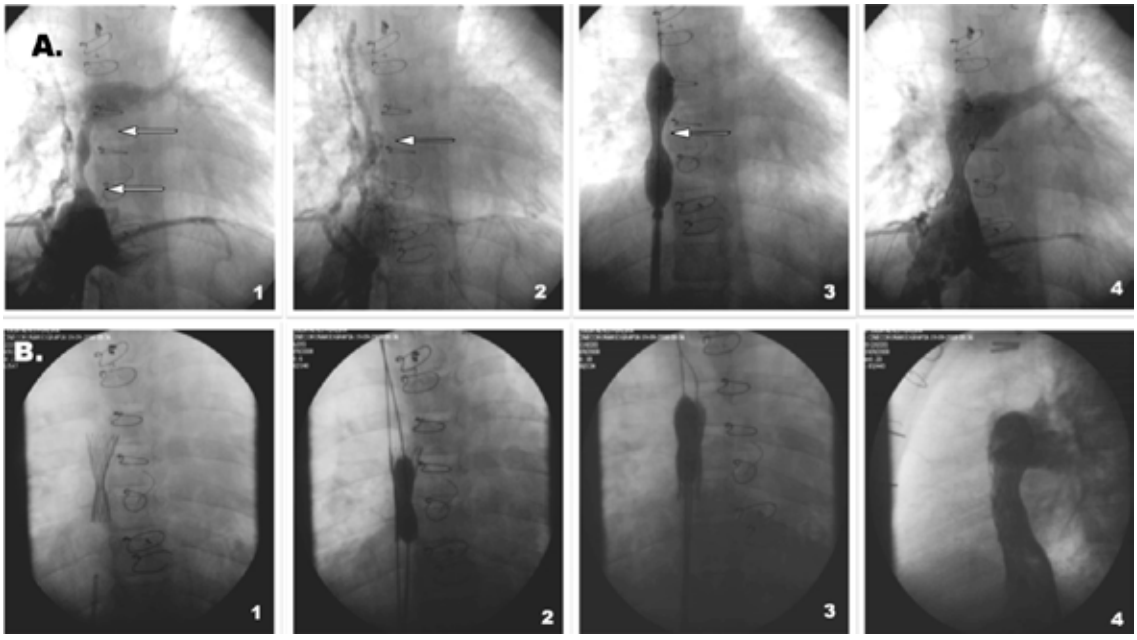

Figure 2 - Angiographic aspects pre A1: diffuse stenosis on ECC (arrows), A2: collateral veins (arrow); A3: balloon inflated with limit rupture pressure, keeping stenosis on the tube (arrow), A4: final aspect. B1: poor stent expansion; B2: dilatation with a balloon; B3: dilatation of residual stenosis with two simultaneous high-pressure balloons. B4: final outcome.

in the development of ascites, a common clinical element among patients.

In case 1, the Fontan physiology was established by a Gore-Tex ECC, and a moderate stenosis at the anastomosis level with the inferior vena cava was found in a ten-year child with PLE condition and chronic ascites refractory to medical therapy. The temporary relief of symptoms after stent implantation not only stressed the importance of obstruction in the pathophysiology of clinical manifestations, but also the multifactorial nature of these. In this case, the maintenance of high pulmonary pressures led the surgery team to a second operation, involving the fenestration to a new tube without stenosis. With such measures, there is no need for patient readmissions.

In case 2, an ECC with biological material resulted in a postoperative development with severe events such as pulmonary thromboembolism and severely progressive diffuse luminal narrowing at the tube level. The extreme difficulty of stent expansion suggests the chronic inflammatory nature of the stenosis.

Stanislav et al have recently reported the importance of maintaining surgical conduits patent and without stenosis, recommending percutaneous approach even in cases where the obstruction is angiographically mild with no significant pressure gradient. They underscored the difficulty of detecting gradients in low-pressure system and the fact that these obstructions may impede passive venous flow, particularly in opposed to the gravitational direction[7].

PLE remains a clinical syndrome of unknown etiology. Peripheral edema, ascites and pleuro-pericardial effusions are the usual findings. The development of PLE five years after Fontan surgery is present in about up to 30% of patients[8]. It is related to unfavorable hemodynamics after univentricular correction, such as high venous pressure, low cardiac index, high pulmonary vascular resistance and systolic and diastolic ventricular dysfunction. This syndrome is associated with high mortality, with actuarial survival at five years of 46%. Given the unknown etiology, treatment methods are arbitrary and include drug control measures and invasive procedures such as relief of stenosis across the Fontan circuit[9], occlusion of aorto-pulmonary connections, creation of fenestration and heart transplantation. In case 1, the relief of conduit stenosis resulted in clinical improvement, with remission of ECC only for a period of six months. Then, the association of fenestration also required for pressure decompression of the IVC and to control the ECC for a longer period.

In the cases reported, we had no difficulty in using high-profile devices because the children were older and had less problematic venous access. The greatest and most unexpected difficulty was in the second case, due to the impossibility of adequate expansion of low pressure rupture balloons. Therefore, in a second procedure, we used two simultaneous high-pressure balloons, obtaining then the appropriate lumen, without pressure gradient. Patients remained under oral anticoagulation, and we did neither clinically find, nor did the echocardiographic study reveal, thrombus formation at the level of stents, pulmonary thromboembolism or restenosis

The constant monitoring of patients undergoing Fontan surgery in search of stenoses, distortions and extrinsic compressions should be performed, especially those whose univentricular hemodynamics was performed in non-ideal circumstances, and such abnormalities should be treated early for a good development of these patient in the medium and long term.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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Study Association

This study is not associated with any post-graduation program.

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