Journal of
Transcatheter Interventions
e-ISSN e-2595-4350

Angiographic progression of a large pulmonary arteriovenous fistula resulting from Fontan operation: therapeutic implications

Evolução angiográfica de grande fístula arteriovenosa pulmonar decorrente de cirurgia de Fontan: implicações terapêuticas

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DOI: 10.31160/JOTCI201927A20190009

ABSTRACT – Cavopulmonary shunts are the surgical procedures of choice for the correction of congenital heart diseases with univentricular physiology. Unfortunately, its beneficial effects are limited by the development of complications, notably the formation of pulmonary arteriovenous fistulas, which are related to the interruption of suprahepatic venous flow directed to the pulmonary circulation.

Keywords: Arteriovenous fistula; Fontan procedure; Heart defects, congenital; Pulmonary artery/abnormalities

RESUMO – As derivações cavopulmonares são os procedimentos cirúrgicos de eleição para correção de cardiopatias congênitas com fisiologia univentricular. Infelizmente seus efeitos benéficos são limitados pelo desenvolvimento de complicações, notadamente a formação de fístulas arteriovenosas pulmonares, as quais estão relacionadas com a interrupção do fluxo venoso supra-hepático direcionado para a circulação pulmonar.

Descritores: Fístula arteriovenosa; Técnica de Fontan; Cardiopatias congênitas; Artéria pulmonar

How to cite this article:
Chierice JR, Pavão RB, Badran AV, Schmidt A, Manso PH, Marin-Neto JA, et al. Angiographic progression of a large pulmonary arteriovenous fistula resulting from Fontan operation. Therapeutic implications. J Transcat Interven. 2019;27:eA20190009. https://doi.org/10.31160/JOTCI201927A20190009

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Received on:
May 13, 2019

Accepted on:
Aug 27, 2019

INTRODUCTION

Cavopulmonary shunts are the surgical procedures of choice for management of congenital heart diseases in patients with functionally univentricular hearts, and are associated with improved quality of life and longer survival.[1] The Glenn procedure promotes the formation of pulmonary arteriovenous fistulae (PAVF), leading to progressive systemic arterial unsaturation, polycythemia and increased risk of thrombosis.[2]

Restoration of suprahepatic venous drainage into the pulmonary circulation via the Fontan procedure is thought to induce the resolution of PAVF, which are often multiple and small.[3]

This article describes findings of sequential hemodynamic and angiographic studies of a patient followed for 7 years, prior to and after the Fontan operation, demonstrating the resolution of a large arteriovenous fistula in the middle lobe of the right lung. This research was approved by the Hospital das Clínicas of Faculdade de Medicina de Ribeirão Preto of Universidade de São Paulo (protocol 3.500.684, CAAE 18655119.9.0000.5440).

CASE REPORT

Male patient presenting with cyanosis from birth, diagnosed with mitral and pulmonary atresia by echocardiography performed at a hospital located at the pa-

tient's city of origin, and submitted to systemic-pulmonary anastomosis in the first month of life at the same hospital. At the age of 2 years, the child was referred to our organization, where the correct echocardiographic diagnosis of *situs solitus*, ventricular inversion, univentricular atrioventricular connection associated with tricuspid atresia and ventriculo-arterial discordance with ventricular and atrial septal defects was given. At the age of 2 years and 3 months, surgical ligation of the systemic-pulmonary anastomosis and pulmonary artery banding were performed, followed by pulmonary artery transection and bidirectional Glenn shunt 6 months later. Pre-Fontan hemodynamic assessment was carried out within 6 months of the Glenn operation, and revealed pulmonary artery pressure of 90×55 (66) mmHg and pulmonary vascular resistance of 7.04UW/m^2, precluding the performance of the total cavopulmonary shunt procedure. The patient was then treated with sildenafil and scheduled for reassessment within 6 months. However, the family did not bring him for follow-up visits until 3 years and 11 months later, when mean pulmonary pressure consistent with performance of a total cavopulmonary shunt (15mmHg) was recorded. The fenestrated Fontan procedure was performed; however, severe hemodynamic instability with systemic arterial hypotension and drop in oxygen saturation from 88% to 66% occurred 9 days later. Cardiac catheterization revealed tight stenosis of the inferior vena cava to extracardiac tube graft anastomosis, patent fenestration, multiple micro PAVF in both lungs, and a large fistula in the middle lobe of the right lung (Figure 1). Anastomotic stenosis dilation was achieved with a 39-mm Andra stent (Andramed®) (Figure 2), increasing systemic arterial oxygen saturation to 81% and reversing arterial hypotension.

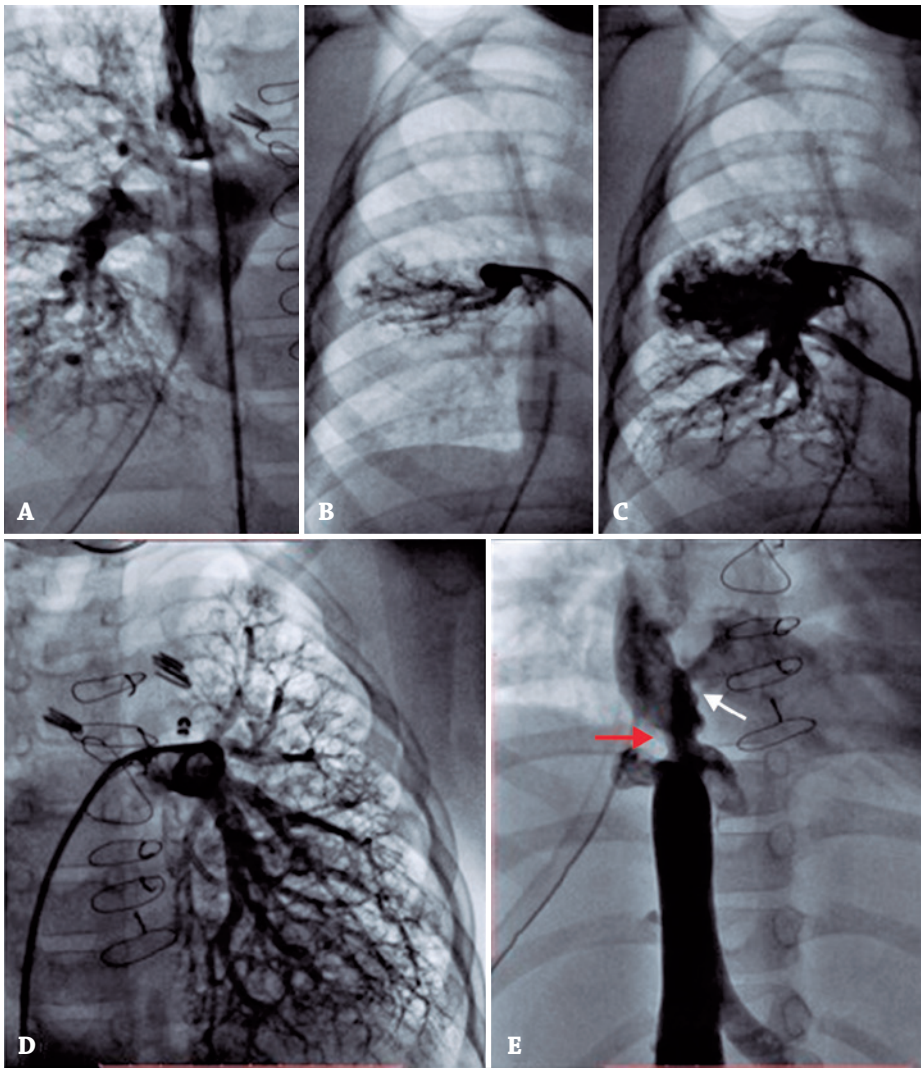

Figure 1. Angiography 3 years after Glenn and 9 days after Fontan. (A) Injection in the superior vena cava demonstrating micro pulmonary arteriovenous fistulas in the right lung. (B and C) Sequential images of selective injection in the middle pulmonary artery of the lung evidencing large pulmonary arteriovenous fistula. (D) Micro pulmonary arteriovenous fistulas in the left lung. (E) Obstruction of the extra-cardiac tube anastomosis with the inferior vena cava (red arrow) and fenestration of the extra-cardiac tube (white arrow).

Progressive drop in arterial oxygen saturation (from 81% to 70%) and polycythemia with hemoglobin concentration of 18.1g/dL developed over the course of the next 3 years, when he was followed up at the outpatients clinic. Echocardiography revealed persistent fenestration and percutaneous occlusion was therefore indicated. During this procedure, selective arteriograms of both pulmonary branches confirmed PAVF resolution, including the large right lobe fistula (Figure 3). Fenestration occlusion was achieved with a 5×6mm Amplatzer duct occluder (Amplatzer Duct Occluder II, GA Medical Corporation®). Control angiography revealed lack of residual flow (Figure 4) and arterial oxygen saturation increased to 92%.

Non-implantation of occlusion devices at the time of initial diagnosis, and temporary loss to follow-up (i.e., non-attendance to scheduled follow-up visits) allowed the documentation of development and subsequent resolution even of large PAVF following the Glenn and Fontan procedures.

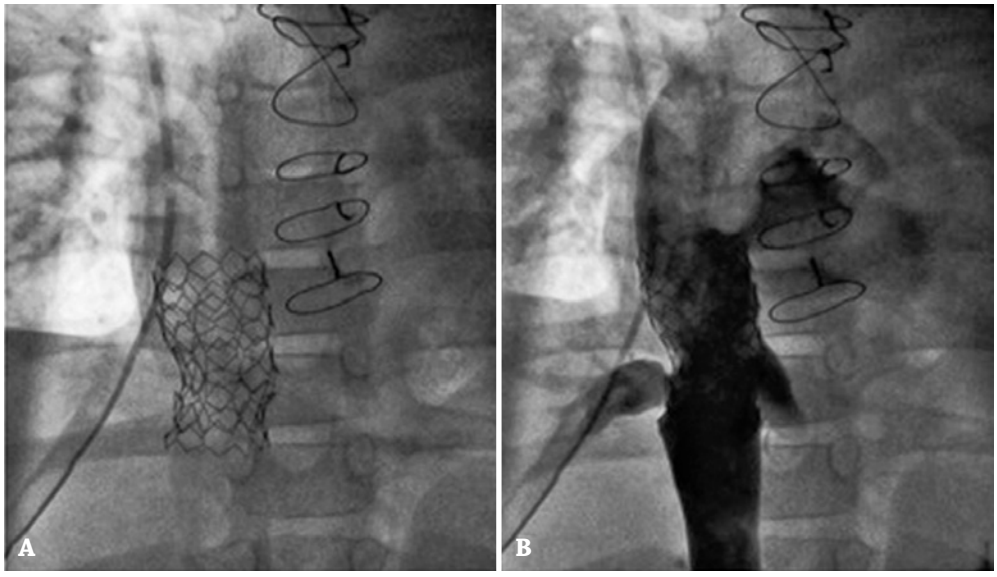

Figure 2. Dilatation of the obstruction in the anastomosis of the extra-cardiac tube with the inferior vena cava. (A) Implanted stent. (B) Control angiography.

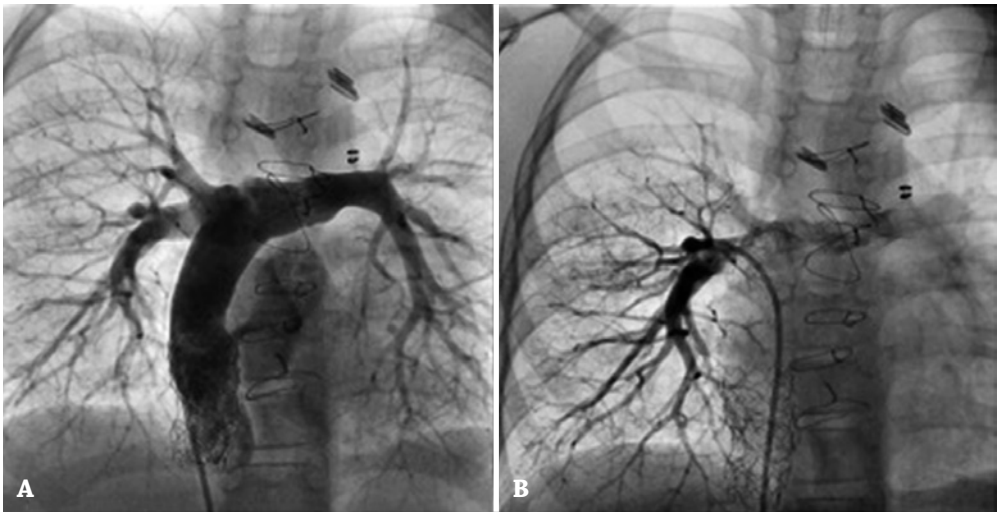

Figure 3. Evaluation of pulmonary arteriovenous fistulas prior to fenestration occlusion. (A) Extra-cardiac tube angiography demonstrating absence of pulmonary arteriovenous fistulas in both lungs. (B) Selective arteriography of the lower and middle lobe showing absence of the large fistula visible in figure 2.

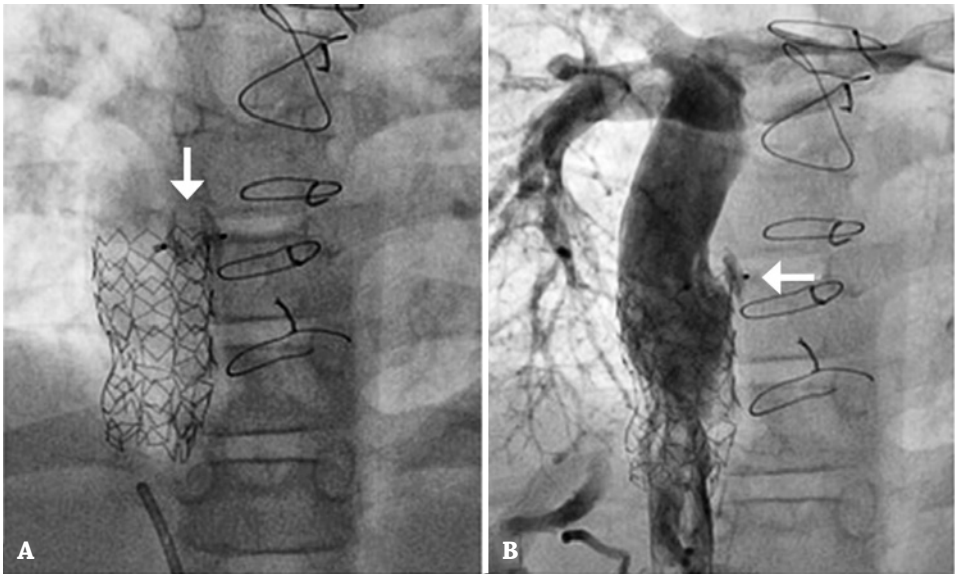

Figure 4. Occlusion of fenestration. (A) Amplatzer Duct Occluder II 5×6 mm (AGA Medical Corporation) (arrow). (B) Control angiography demonstrating no flow at the prosthesis site (arrow).

DISCUSSION

Glenn procedure, a step in staged transition to the Fontan procedure with total cavopulmonary shunt, is thought to be an adequate palliative surgery promoting significant increase in arterial oxygen saturation without the typical volume overload of the systemic ventricle associated with the Blalock-Taussig and similar procedures.[1] Unfortunately, benefits of this procedure are limited by PAVF development and resulting progressive drop in oxygen saturation and polycythemia, among other deleterious effects.[2] The embryological origin of PAVF is related to the persistence and subsequent growth of small communications, which correspond to remnants of the fetal microvascular plexus.[3] Two factors may be considered in the etiopathogenesis of PAVF after the Glenn procedure: (1) the nonpulsatile flow from the venous anastomosis as the only source of pulmonary blood flow[4] – FAVP involution is expected to occur soon after total cavopulmonary shunt operation; however, cases of late FAVP occlusion after the Fontan procedure with or without prior Glenn procedure have been reported;[5] and (2) lack of hepatic anti-angiogenic factors and pulmonary circulation vasodilation inhibitors potentially associated with imbalances between angiogenic and vasodilating factors.[6,7] Scintigraphy studies revealed the development of PAVF in virtually all patients undergoing bidirectional Glenn operation.[8]

Redirection of the hepatic venous flow to the pulmonary circulation is thought to induce PAVF resolution.[2] The case described confirms this resolution and emphasizes the role of hepatic factors in the genesis of vascular malformations after the Glenn procedure.

Total right heart bypass, often achieved via the Fontan operation, is an effective surgical procedure for correction of several complex congenital heart diseases, allowing the restablishment of in-series systemic-pulmonary circulations associated with univentricular hearts. Current perioperative morbidity and mortality rates associated with the Fontan procedure are low; however, chronically elevated systemic venous pressure has negative impacts on several organs in the mid-term. Late pathological changes, such as protein-losing enteropathy, persistent pleural effusion, chronic liver disease, arrhythmias and plastic bronchitis, result in high morbidity and mortality.

Udeken et al.[9] reported loss of post-Fontan surgery beneficial effects within 15, 20 and 25 years in 17%, 30% and 44% of patients respectively, with progression to death or referral for heart transplantation as final outcomes. Elder et al.[10] reported a 59.8% incidence of patients not progressing to mortality or cardiac transplantation within 30 years. Most patients experiencing Fontan failure due to severe pathologic changes in multiple organs are not eligible for cardiac transplantation, and progress to death in the short term.[11]

Pulmonary arteriovenous fistula occlusion in patients submitted to the Glenn procedure should be limited to cases with marked arterial oxygen unsaturation, since these fistulae tend to resolve spontaneously after total cavopulmonary bypass surgery, with return of oxygen saturation to values consistent with this condition.

Mid- to long-term effects of the Glenn procedure often prevent pathological changes in multiple organs resulting from elevated systemic venous pressure after the Fontan procedure. This management strategy may also allow timely cardiac transplantation to overcome the ventricular failure

that tends to supervene in the third or fourth decade of life, decreasing the mortality rates commonly associated with this procedure. Development of PAVF with significant cyanosis in the mid-term after a Glenn procedure is the major limiting factor of this management plan. The occurrence of PAVF and related deleterious consequences are therefore robust indications for total cavopulmonary shunt. Deeper understanding of hepatic factors associated with angiogenesis regulation and scientific advances in their pharmacological modulation may help prevent PAVF formation, so as to allow partial cavopulmonary shunts to be maintained as an effective palliative therapy alternative for many years.

SOURCE OF FINANCING

None.

CONFLICTS OF INTEREST

The authors declare there are no conflicts of interest.

CONTRIBUTION OF AUTHORS

Conception and design of the study: JRAC and JLH; coleta dos dados: JRAC; data collection: João Reynaldo Abbud Chierice; data interpretation: JRAC, RBP, AVB, PHM and JLH; text writing and approval of the final version to be published: JRAC, JLH and JAMN.

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