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

Interventional therapeutic procedures in cavopulmonary shunts

Procedimentos terapêuticos intervencionistas em derivações cavopulmonares

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How to cite this article:
Oliveira RM, Haddad JL, Pavão RB, Badran AV, Figueiredo GL, Lago IM, et al. Interventional therapeutic procedures in cavopulmonary shunts. J Transcat Interven. 2018;26:eA0009.https://doi.org/10.31160/JOTCI2018;26(1)A0009

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Submitted on:
Oct 3, 2017

Accepted on:
Jul 25, 2018

DOI: 10.31160/JOTCI2018;26(1)A0009

ABSTRACT – **Background** Cavopulmonary shunts are the palliative surgeries of choice in congenital heart diseases with the anatomical and pathophysiological defect of univentricular heart. The objective of this study was to report the results of percutaneous procedures in patients with complications following partial or total surgical cavopulmonary shunts. **Methods**: The procedures were conducted under general anesthesia and continuous monitoring of hemodynamic and respiratory parameters. The accesses used were the femoral vein (Fontan) or the internal jugular vein (Glenn). The devices were those commonly used for treating vascular obstructions, occlusion of venovenous or arteriovenous shunts, and systemic-pulmonary anastomoses. **Results**: Ten procedures were conducted in ten patients; in that, four Glenn (Group I) and six Fontan (Group II). The mean age in Groups I and II was 86 months and 106 months, and the mean time since surgery was 24 months and 25 months, respectively. Six patients had obstructive lesions and four had venovenous shunts, systemic-pulmonary anastomosis or a fenestrated cavopulmonary tube. In cases with obstructive lesions, there was no residual gradient; after dilation, there was a significant increase or normalization of vessel diameter. In patients with shunts, there was complete occlusion after deployment of the device. In all cases hemodynamic parameters normalized, clinical status improved and there were no complications. **Conclusions:** Percutaneous procedures are safe and effective to treat obstructive lesions or shunts in patients with a history of cavopulmonary shunt and presenting such complications.

Keywords: Fontan procedure; Stents; Prostheses and implants; Heart defects, congenital

RESUMO – **Introdução:** As derivações cavopulmonares constituem cirurgias paliativas de eleição em cardiopatias congênitas com condição anatomofisiopatológica univentricular. Este estudo objetivou relatar os resultados dos procedimentos percutâneos em pacientes com complicações após cirurgias de derivação cavopulmonar parcial ou total. **Métodos:** Os procedimentos foram realizados sob anestesia geral e monitorização contínua dos parâmetros hemodinâmicos e respiratórios. A via de acesso foi obtida por punção da veia femoral (Fontan) ou da veia jugular interna (Glenn). Os dispositivos utilizados foram os de uso habitual para o tratamento de obstruções vasculares, de oclusão de *shunts* venovenosos e arteriovenosos, e das anastomoses sistêmico-pulmonares. **Resultados:** Foram realizados dez procedimentos em dez pacientes, sendo quatro com cirurgia de Glenn (Grupo I) e seis com cirurgia de Fontan (Grupo II). A média de idade foi de 86 e 106 meses, e o tempo médio decorrido após a cirurgia foi de 24 e 25 meses nos Grupos I e II, respectivamente. Seis pacientes apresentavam lesões obstrutivas e quatro, *shunts* venovenosos, anastomose sistêmico-pulmonar ou fenestração do tubo cavopulmonar. Nos casos com lesões obstrutivas, houve ausência de gradiente residual e, após a dilatação, aumento significativo ou normalização do diâmetro do vaso. Nos pacientes com *shunts*, houve oclusão total após o implante do dispositivo. Em todos os casos, houve normalização dos parâmetros hemodinâmicos, melhora no quadro clínico e ausência de complicações. **Conclusões:** Os procedimentos terapêuticos percutâneos são seguros e eficazes no tratamento das lesões obstrutivas ou na oclusão de *shunts* em pacientes submetidos à derivações cavopulmonares, que apresentam tais complicações em sua evolução.

Descritores: Técnica de Fontan; Stents; Próteses e implantes; Cardiopatias congênitas

BACKGROUND

Cavopulmonary shunts (CPS) are the palliative surgeries of choice in congenital heart diseases with the anatomical and pathophysiological univentricular hearts. Partial CPS, or modified, or bidirectional Glenn procedure redirects the flow of the superior vena cava to the pulmonary branches through an end-to-side anastomosis with the right branch of the pulmonary artery.[1,2] In a second surgery, the flow of the inferior vena cava is diverted by the interposition of a tube between this vein and the right pulmonary artery, completing the total shunt of the systemic venous flow, without a ventricular chamber (modified Fontan procedure).[3-5]

Several anatomical complications can occur in the follow-up of patients that have undergone CPS, which affect the delicate balance of a circulation that has little reserve to cope with pressure or volume overloads.[6-10] The percutaneous treatment of obstructive lesions or deleterious shunts, considering the use of regular devices, is an essential resource to restore appropriate physiological conditions, therefore improving quality of life, prolonging survival, and either avoiding new surgical interventions or enabling them to be conducted under more favorable clinical conditions.

The objective of this study was to report the results of percutaneous procedures in patients with complications following partial or total surgical cavopulmonary shunt.

METHODS

This is a retrospective and observational study of CPS patients that have undergone percutaneous therapeutic procedures at the *Hospital das Clínicas da Faculdade de Medicina de Ribeirão Preto da Universidade de São Paulo* (USP). The present study was submitted to the Research Ethics Committee of the Hospital das Clínicas of the Medical School of Ribeirão Preto of USP (CAAE No. 98346718.6.0000.5440) and was approved on September 24, 2018.

The procedures were conducted under general anesthesia with orotracheal intubation and continuous monitoring of hemodynamic and respiratory parameters. The arterial access was performed in all cases by femoral approach, and the venous access was obtained by ultrasound-guided internal jugular vein puncture in Glenn procedure patients, and by femoral vein puncture in Fontan operation patients. Patients received anticoagulation with unfractionated heparin at a dose of 100IU/kg body weight.

The anatomical complications were evaluated before and after the procedures by angiography in different projections, according to their location, complemented by gradient determination in cases of obstruction, and saturation evaluation in cases of shunts. The obstructions were treated with stent placement. Run-off veins, systemic-pulmonary anastomoses, and venovenous fistulas were occluded with vascular plugs, and fenestration was occluded with atrial septal occluder.

RESULTS

In the period from January 2012 to December 2016, ten patients underwent ten percutaneous therapeutic procedures with equal sex distribution. Group I comprised four bidirectional Glenn procedure patients (40%), and Group II encompassed six Fontan operation patients (60%). Mean age of patients was 86 (12 to 192) months and 106 (56 to 288) months, and the mean time since surgery was 24 (zero to 264) months and 25 (2 to 72) months in Groups I and II, respectively. The procedures performed are listed in Table 1. In all cases, there was immediate success with improvement of hemodynamic conditions and no complications.

Table 1. Interventional percutaneous procedures performed

Patient	Age (months)	Sex	Surgery	Time after surgery	Lesion	Procedure	Pre sat%	Post sat%	Result
1	288	Female	Fontan	22 years	Stenosis of the RBPA	Angioplasty + stent	82	92	No stenosis
2	82	Female	Fontan	1 day	Stenosis of the RBPA	Angioplasty + stent	72	82	No stenosis
3	113	Female	Glenn	6 days	Stenosis of the LBPA	Angioplasty + stent	63	85	Nostenosis
4	64	Male	Fontan	9 days	Tube obstruction	Angioplasty + stent	64	73	No stenosis
5	62	Male	Fontan	35 days	Embolism in the LBPA	Recanalization with baloon	89	95	Better perfusion
6	56	Male	Fontan	5 days	Trombosis of the superior vena cava	Balloon angioplasty	82	91	No stenosis
7	192	Male	Glenn	2 years	Venovenous fistula	Plug occlusion	90	98	Absence of flow
8	28	Female	Glenn	2 months	Runoff vein	Plug occlusion	75	82	Absence of flow
9	12	Male	Glenn	3 months	Systemic-pulmonary anastomoses	Plug occlusion + coil	70	85	Absence of flow
10	84	Female	Fontan	3 years	Tube fenestration (5mm)	Occlusion of the graft	80	92	Absence of flow

Sat%: systemic saturation (before and after the procedure); RBPA: right branch of the pulmonary artery; LBPA: left branch of the pulmonary artery.

Stents were placed in four patients - three in obstructions of pulmonary branches and one in the anastomosis of the tube with the inferior vena cava. One patient had undergone a classic Fontan procedure and presented with stenosis of the right branch of the pulmonary artery due to aneurysm dilation in the right atrium. Surgical correction was contraindicated due to protein-losing enteropathy and chronic liver disease (Figure 1). In another case with anastomotic stenosis of the tube to the inferior vena cava, the procedure was conducted in the immediate postoperative period (Figure 2). The patient had 75% stenosis with a gradient of 8mmHg and was treated with a 30-mm Andra XL® stent (Andramed, Reutlingen, Germany). After deployment, there was no gradient. However, there was a residual angiographic stenosis of 20%. We decided not to redilate the stent with a high-pressure balloon because of recent suture and absence of a residual gradient. In the remaining patients with stenosis of pulmonary branches, Palmaz® Genesis™ stents (Cordis, Baar, Switzerland) were used, resulting in absence of gradient and residual angiographic stenosis.

Balloon angioplasty was perfomed in two patients with thrombus obstruction, located in the left branch of the pulmonary artery, and in the anastomosis of the inferior vena cava with the extracardiac tube. Systemic saturation and pulmonary flow improved in both cases.

Two patients underwent venous occlusion with CERA™ vascular plug (Lifetech Scientific Co. Ltd., Shenzhen, China): a run-off vein between the superior and inferior vena cava (Figure 3) and a venovenous fistula between a branch of the innominate vein and the left pulmonary vein (Figure 4). In both patients there was significant systemic desatu-

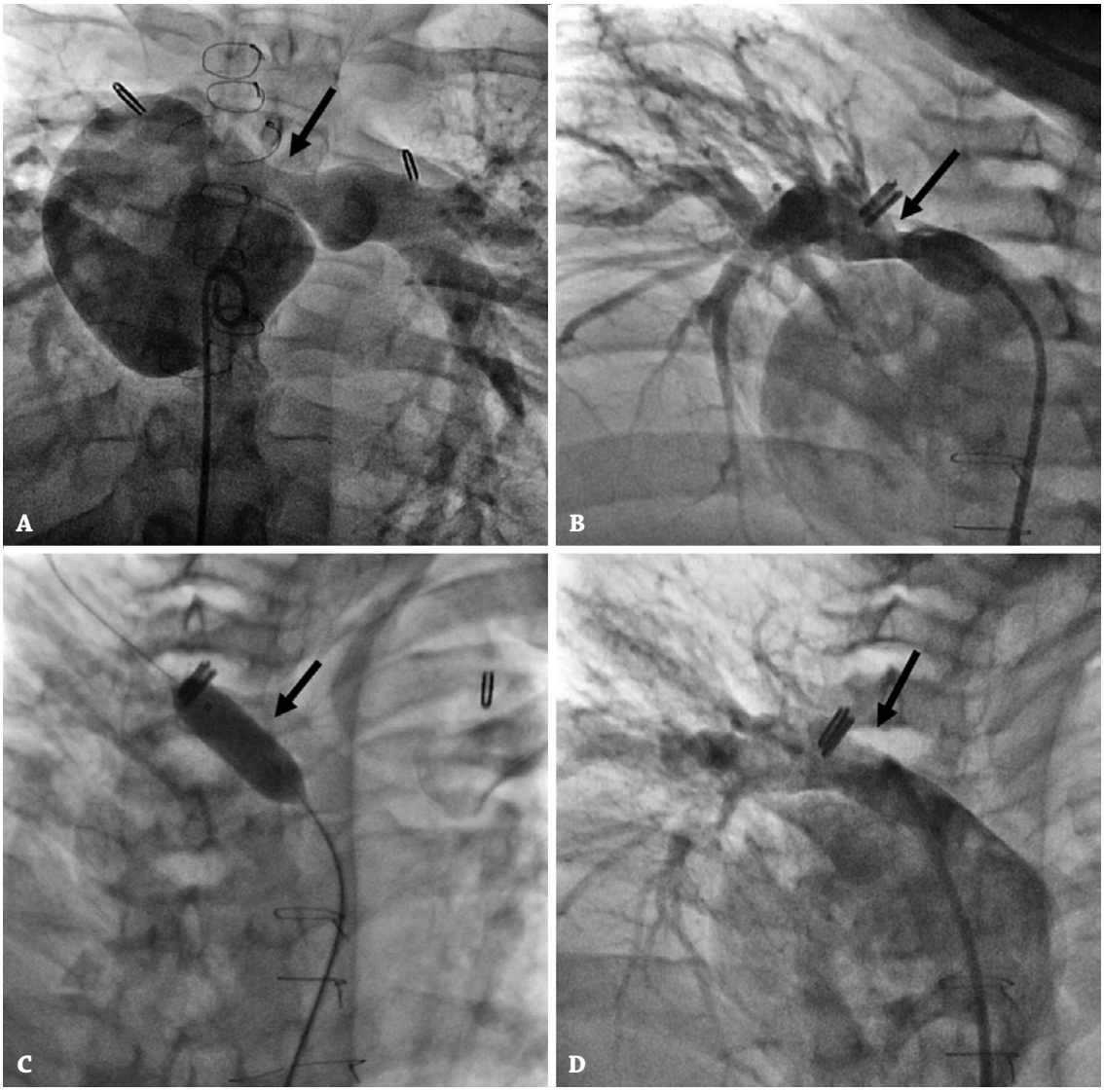

Figure 1. Fontan operation. (A) Aneurysm of the right atrium compressing the right branch of the pulmonary artery (arrow). (B) Selective angiography of right pulmonary branch, showing obstruction before bifurcation (arrow). (C) Stent deployment in the right branch of the pulmonary artery (arrow). (D) Absence of residual stenosis where the stent was placed (arrow).

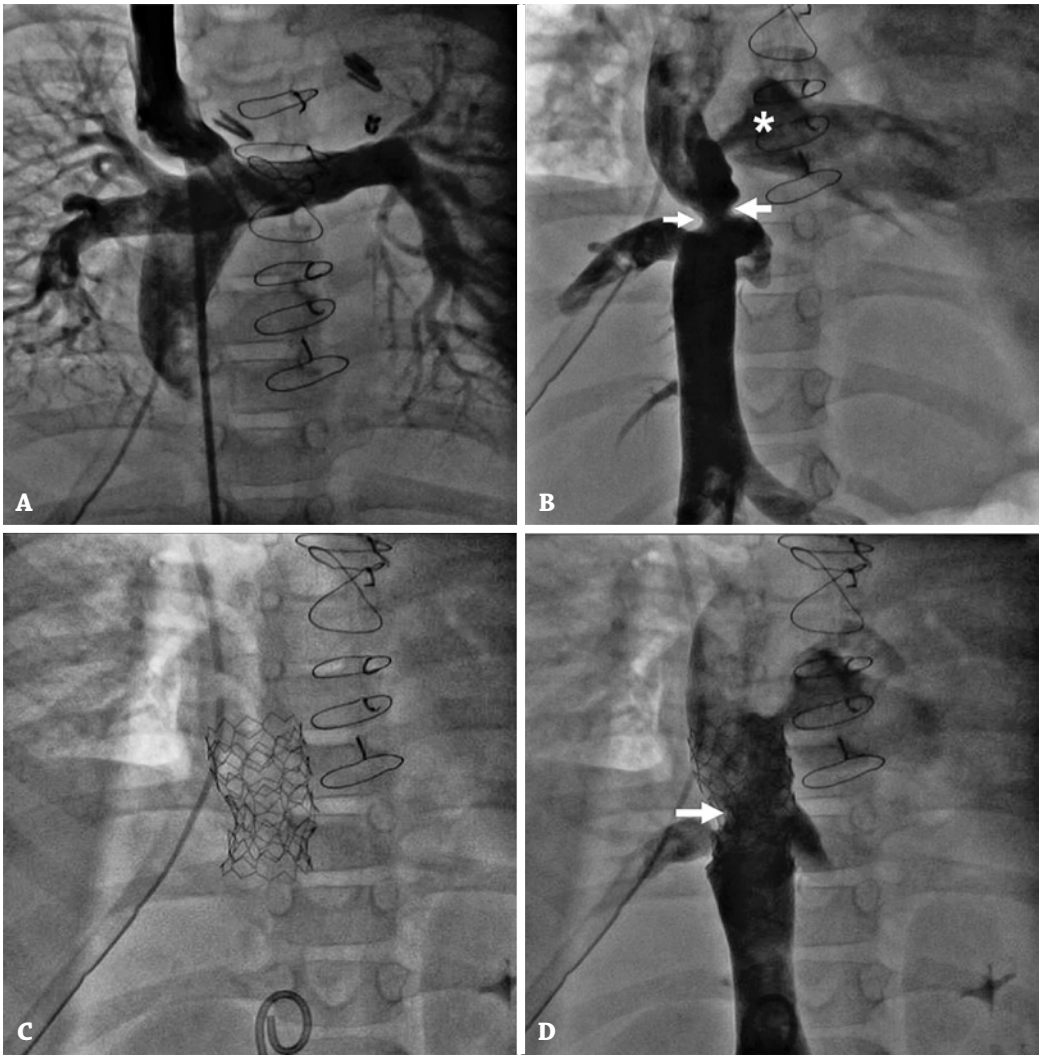

Figure 2. Fenestrated Fontan operation. (A) Angiography of the superior vena cava (Glenn procedure). (B) Angiography of the inferior vena cava showing stenosis of the anastomosis to the extracardiac tube (arrows) and fenestration (asterisk). (C) Stent deployed at the site of anastomosis. (D) Control angiography showing a small residual lesion.

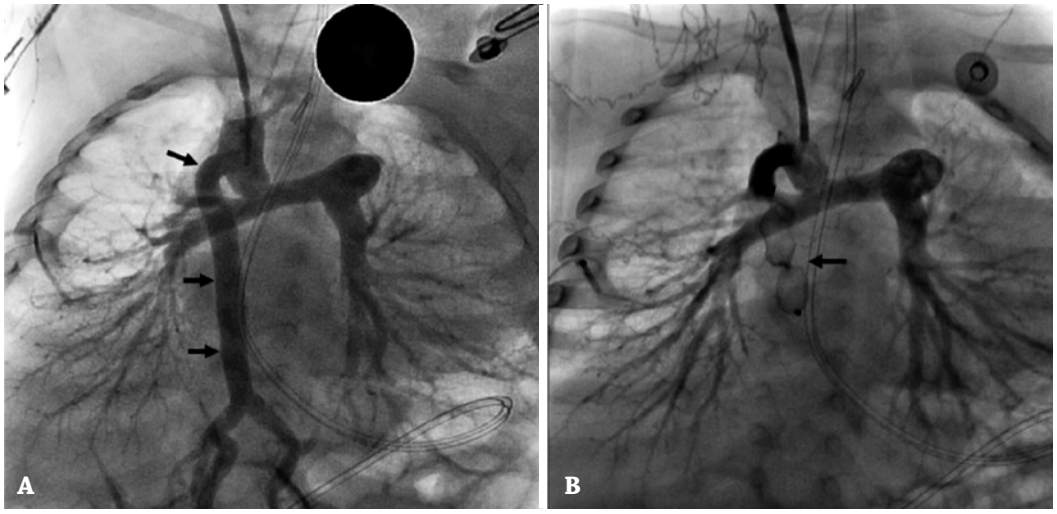

Figure 3. Bidirectional Glenn procedure. (A) Large run-off vein from the superior to the inferior vena cava. (B) Selective angiography of the run-off vein, showing vascular plug with total occlusion (arrow).

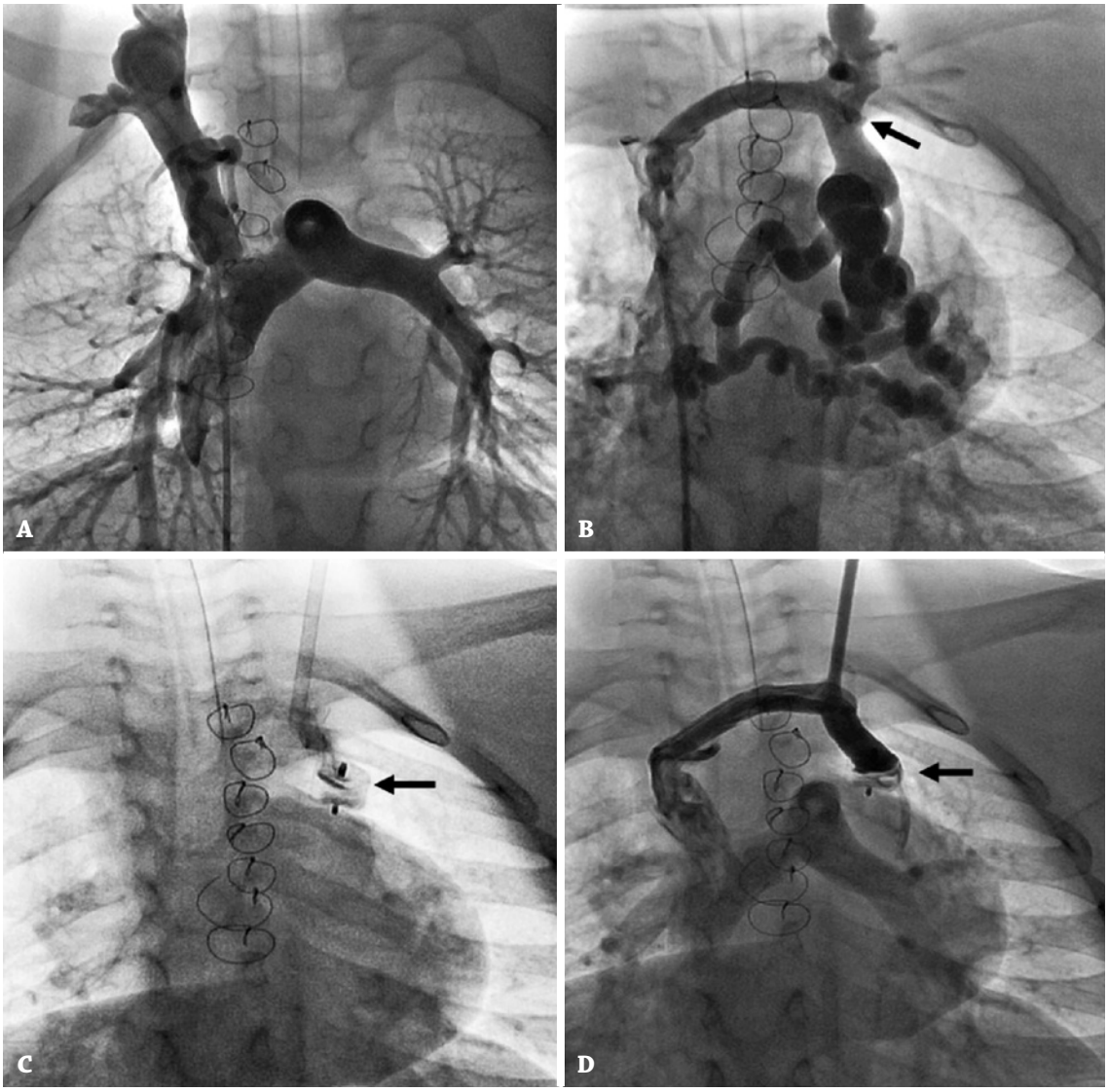

Figure 4. Modified Fontan operation. (A) Selective angiography of the superior vena cava, demonstrating anastomosis with no obstruction. (B) Large vein originating from the confluence of the left jugular vein with the inominate vein (arrow), with multiple fistulas for the pulmonary veins. (C) Vascular plug deployed in the initial segment of the fistulous vein (arrow). (D) Selective angiography in the segment proximal to the plug showing total occlusion.

ration consequent to the reduction of pulmonary flow, the veins being completely occluded. After the Glenn procedure, there was subsequent increase in saturation back to normal values.

One patient with Glenn procedure had two systemic-pulmonary anastomoses, originating in the descending aorta to both branches of the pulmonary artery (Figure 5). The anastomosis to the right branch was large, with high flow, and the anastomosis to the left branch had important obstructions (one at the aortic ostium, and another from the middle segment to the connection with the pulmonary artery). The first was occluded with CERA™ vascular plug and the second with a Gianturco® coil (Cook Medical Inc., Bloomington, USA).

A persistent 5-mm fenestration in a patient who presented systemic saturation of 80% was occluded with CERA™ ASD Occluder (6mm) (Figure 6). The result was absence of residual shunt and increase in systemic saturation to 92%.

DISCUSSION

Fontan operation was described in 1971, and consists of an anastomosis between the right atrium and the pulmonary artery. It enabled establishing serial systemic-pulmonary circulation, excluding the pump function of the ventricle at the origin of the pulmonary flow, in cases of congenital heart defects with anatomically or functionally univentricular heart. The surgical technique of this diver-

Figure 5. Glenn procedure. (A and B) Selective angiographies in the right and left branches of the pulmonary artery. (C) Important systemic-pulmonary anastomosis for the right branch, originating from the descending aorta (arrow). (D) Total occlusion of the systemic -pulmonary anastomosis to the right branch with a vascular plug and coil (arrows). (E) Stenotic pulmonary systemic anastomosis (arrow) to the left branch. (F) Total occlusion of the systemic-pulmonary anastomosis to the left branch (arrow).

sion of the systemic venous return to the pulmonary artery had several modifications over the years. Currently, serial circulation is obtained in two stages: first an end-to-side anastomosis of the superior vena cava to the right branch of the pulmonary artery (modified Glenn procedure); in the second stage, the interposition of a tube between the inferior vena cava and the pulmonary artery achieves complete diversion of the systemic venous flow.[1-5]

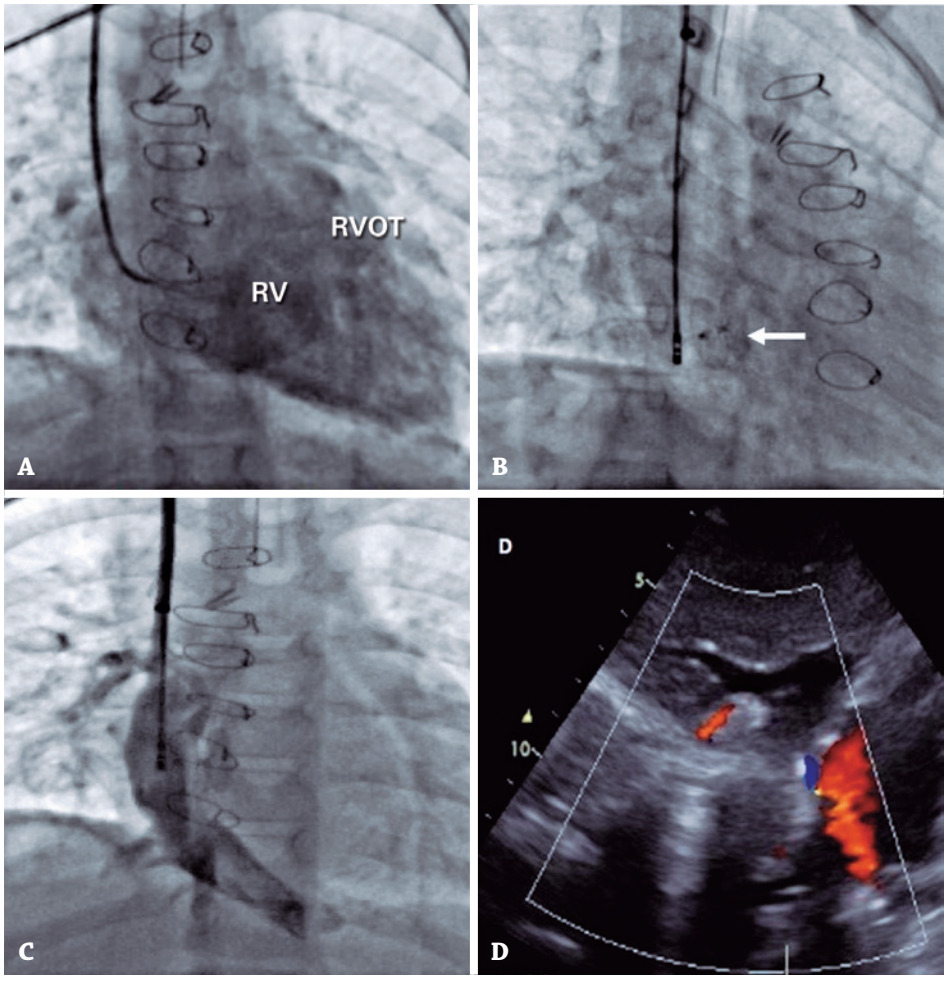

Figure 6. Fontan Operation. (A) Manual injection at the fenestration, demonstrating opacification of the right ventricle (RV) and of the right ventricle outflow tract (RVOT). (B) Occluder of interatrial communication deployed at the fenestration (arrow). (C) Control angiography of the tube between the inferior vena cava and the right branch of the pulmonary artery, showing minimal residual shunt through the mesh. (D) Follow-up Color Doppler one month after the occlusion, demonstrating absence of flow from the tube to the left atrium.

Most patients with univentricular heart require a palliative procedure prior to Glenn operation. Cerclage of the pulmonary artery is indicated in cases of excess pulmonary flow, to avoid pulmonary hypertension incompatible with the CPS. In cases of pronounced low pulmonary flow, a systemic-pulmonary shunt should be created to obtain adequate systemic arterial saturation (for example, modified Blalock-Taussig surgery). These procedures may also cause distortion of the pulmonary branches and obstructions in the postoperative period.

In patients with congenital cardiac diseases and univentricular heart that have undergone CPS, postoperative complications are common, such as stenoses of pulmonary branches and of anastomotic sutures, thromboses, persistence of fenestration or of systemic-pulmonary anastomoses, presence of run-off veins, venovenous fistulas, chylothorax, plastic bronchitis, protein-losing enteropathy and chronic liver disease.[11-15]

Structural malformations cause increased venous pressure, modifying the complex pathophysiology of the univentricular circulation. In these situations, interventional therapeutic procedures may restore circulatory balance and improve clinical conditions, therefore avoiding or, at least, postponing another surgical intervention.[16-20] The procedures in this series were simple, using devices usually available in the interventional arsenal. In obstruction secondary to stenosis in the suture of the tube to the vena cava, dilation was conducted in the immediate postoperative period. The obstructions at the anastomotic suture lines can be approached early, as long as they have been made with running sutures, which allows, through dilation, an increase in the circumference of the suture. Fenestration of the anastomotic tube is not usually performed in our institution; therefore, only one case required occlusion of the fenestration due to important systemic desaturation.

Our study has limitations. The series is modest, there is no long-term follow-up to demonstrate late efficacy of the procedures, maintaining the immediate results obtained. The department is considered of medium size and the CPS

program is recent. The results are, however, comparable to those of large series published in the literature.[20-23]

CONCLUSIONS

Percutaneous treatment of obstructive complications and modifications in pulmonary flow secondary to fenestrations, fistulas, or persistence of systemic-pulmonary anastomoses is the procedure of choice in situations that affect the complex physiology of cavopulmonary shunts. These procedures are safe, the technique is predictable and the devices used are commonly available, providing immediate satisfactory results.

FINANCING

None.

CONFLICTS OF INTEREST

The authors declare no conflict of interest.

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