Cardiology in the Young

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Balloon angioplasty of bidirectional Glenn anastomosis

Amr Matoq and Wolfgang Radtke

Nemours Cardiac Center, Alfred I. duPont Hospital for Children, Wilmington, DE 19803, USA

Original Article

Cite this article: Matoq A and Radtke W (2020) Balloon angioplasty of bidirectional Glenn anastomosis. *Cardiology in the Young* **30**: 1452–1457. doi: 10.1017/S1047951120002292

Received: 20 November 2019
Revised: 15 May 2020
Accepted: 12 July 2020
First published online: 11 August 2020

Keywords:
Single ventricle; veno-venous collaterals; stenosis

Author for correspondence:
Wolfgang Radtke MD, Nemours Cardiac Center, Nemours/Alfred I. duPont Hospital for Children, 1600 Rockland Rd, Wilmington, DE 19803, USA. Tel: +1 302 651 6600; Fax: +1 302 651 6600. E-mail: Wolfgang.Radtke@nemours.org

Abstract

Objective: We aim to assess the safety and efficacy of the transcatheter balloon dilation of superior cavopulmonary anastomosis (SCPA). *Background:* SCPA stenosis can lead to impaired pulmonary blood flow, hypoxemia and development of veno-venous collaterals with right-to-left shunt. Balloon dilation of SCPA has been rarely reported and follow-up information is lacking. *Methods:* We performed a retrospective review of patients who underwent cardiac catheterisation and angioplasty of SCPA and reviewed patient's demographics, diagnosis, SCPA surgery and post-operative course, catheterisation haemodynamics, procedural technique, angiography, and the findings of follow-up catheterisation. *Results:* Between 2008 and 2017, seven patients showed significant narrowing of SCPA and underwent balloon angioplasty, all of whom had undergone bidirectional Glenn (BDG). Indications for cardiac catheterisation included persistent pleural effusion, hypoxemia, and echocardiographic evidence of BDG stenosis or routine pre-Fontan assessment. Five patients had bilateral SCPA. The procedure was successful in all cases with increase in the stenosis diameter from a median of 3.3 mm (range 1.2–4.7 mm) to a median of 4.7 mm (range 2.6–7.8 mm). All patients had at least one follow-up cardiac catheterisation. Only one patient required repeat angioplasty at the 2.3-month follow-up with no further recurrence. Sustained results and interval growth were noted in all other cases during up to 29 months of follow-up. No adverse events were encountered. *Conclusion:* Based on our small series, balloon angioplasty of BDG stenosis is feasible and safe and appears to provide sustained improvement with interval growth and only the rare recurrence of stenosis.

The second stage of single-ventricle palliation consists of superior cavopulmonary anastomosis (SCPA), i.e., bidirectional Glenn (BDG) or Hemi-Fontan (HF). Hypoxemia and cyanosis are known complications following the second stage and reported to occur in 6.4% of patients.[1] There are several aetiologies for post-operative hypoxemia including lung disease, low-cardiac output with low mixed venous saturation, and impaired pulmonary blood flow.[2] Stenosis of the SCPA can lead to impaired pulmonary blood flow and hypoxemia. It also promotes development of veno-venous collaterals to the inferior vena cava territory with right-to-left shunt bypassing the pulmonary vasculature. Balloon dilation of SCPA has been rarely reported[3,4] and follow-up information is lacking. In this retrospective study, we aim to assess the safety, efficacy, and clinical outcomes of transcatheter balloon dilation of SCPA.

Materials and methods

At our institution, cardiac catheterisation between SCPA and Fontan completion is performed in all patients. We performed a retrospective chart review of all patients who underwent cardiac catheterisation, following their second-stage palliative surgery, and included all patients with stenosis of the SCPA who underwent catheter intervention for this lesion. We reviewed (1) patient demographics, (2) cardiac diagnosis, (3) SCPA surgical details and post-operative course, (4) cardiac catheterisation haemodynamics, procedural techniques, and angiographic measurements at the time of the index intervention and all subsequent cardiac catheterisations, (5) adverse events following SCPA angioplasty (including access site complication, superior vena cava (SVC) or pulmonary artery (PA) branch tears, or major hemodynamic instability during the procedure), and (6) clinical course before and after SCPA balloon angioplasty. All parameters were entered in Microsoft Excel and mean, median, and range calculated. All cardiac catheterisations were performed under general anaesthesia. Using selective angiograms, SCPA stenosis diameters were measured in perpendicular biplane projections at the junction between SVC and corresponding PA branch (Fig 1). SVC diameter upstream from the stenosis, right and left pulmonary diameter proximal to the first lobar branch, and right and left lower PA branch diameters were measured in perpendicular biplane projections (Fig 1). Balloon sizes were chosen to be at least 2.5 times the stenosis diameter but not larger than 1.6 times the diameter of the adjacent "normal" SVC and PA branch. If the required balloon size was significantly larger than the adjacent PA branches, two balloons side by side were used with one directed

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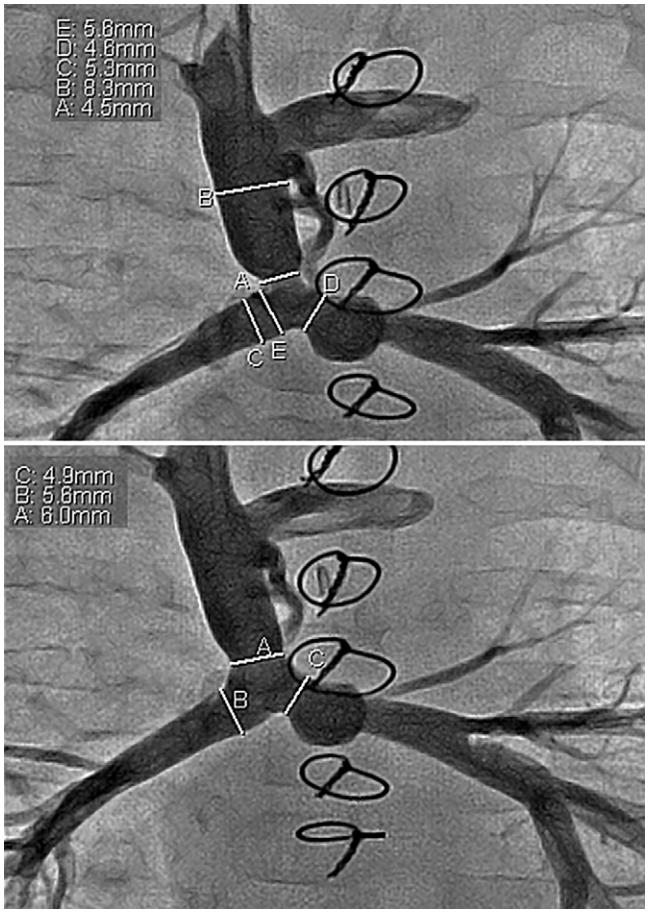

Figure 1. Patient #6; Upper panel showing a pre-angioplasty angiogram in posterior–anterior projection. BDG stenosis (labelled A) and SVC (labelled B) and proximal pulmonary artery (labelled C, D, and E). Lower panel showing a post-angioplasty angiogram, with repeat measurements of BDG and proximal pulmonary artery (A, B, and C respectively).

into the left and one directed into the right branch to achieve the necessary balloon to stenosis ratio (Fig 2). The angioplasty balloons used were NC TREK (Abbott Vascular, California USA), Advance (Cook Medical, Indiana USA), or Sterling (Boston Scientific, Minnesota USA).

Results

Between 2007 and 2017, 90 SCPA procedures were performed in our centre. Of those, 11 (12%) were bilateral SCPA. BDG was performed in only 25 (28%) patients. Of the 90 surgical patients, 84 (93%) patients underwent at least one cardiac catheterisation after second-stage palliation at our institution. Only seven (8%) patients had a significant narrowing of SCPA and underwent balloon angioplasty. No stents were implanted. Patient characteristics, diagnosis, and measurements are summarised in Table 1. Five patients had bilateral SVC; four of them underwent right HF and left BDG, and one patient underwent bilateral BDG. The other two patients underwent a right-sided BDG. The age at SCPA surgery was 3.5–11 months (median 6.1 months), and weight was 5.5–8.4 kg (median 6.6 kg). All patients with SCPA stenosis requiring intervention underwent a BDG and none had a HF at

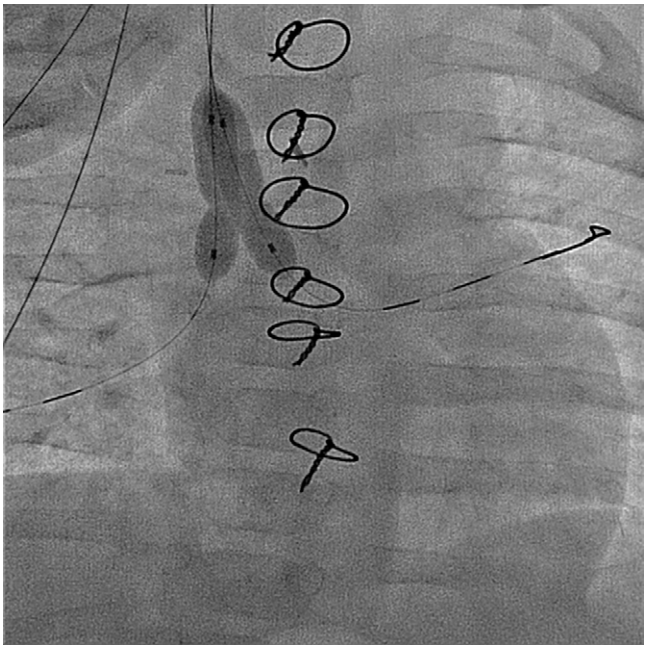

Figure 2. Patient#6, Use of two sides by side balloons to achieve the necessary balloon-to-stenosis ratio despite small pulmonary artery branches.

the site of stenosis. Echocardiography detected the stenosis in four of seven patients.

Cardiac catheterisation

Indications for cardiac catheterisation included persistent pleural effusion, hypoxemia consistently below 80%, or clinical oxygen requirement or suspected BDG stenosis by echocardiography or routine pre-Fontan evaluation. Two patients were noted to have BDG stenosis in the routine pre-Fontan cardiac catheterisation. The stenotic BDG was left sided in four patients and right sided in three patients. BDG angioplasty was performed at a median of 2.8 months (6 weeks–23 months), following SCPA surgery with a median patient weight of 9.2 kg (range 7.7–13.7 kg). The pressure gradient between SVC and PA was relatively low at 2 mmHg mean (range 1–5 mmHg) as expected in the venous system and in the presence of patent azygos veins. Median Qp:Qs was 0.5 (0.45–0.5) pre-angioplasty and had increased to a median Qp:Qs of 0.6 (0.4–0.8) at follow-up cardiac catheterisation. Six of the seven patients did not have ligation of the azygos vein at the time of SCPA surgery, as it is the routine in our centre. All of those patients had run-off via the azygos vein at the time of cardiac catheterisation, resulting in oxygen saturations in the catheterisation laboratory of a median of 75% (65–85%), which increased to a median of 80% (77–89%) as an expression of decreased veno-venous collateral run-off.

Balloon angioplasty

Access to the stenotic BDG was obtained from the ipsilateral internal jugular vein (Fig 3) in four patients and from the contralateral internal jugular vein (Fig 4) in three patients. Balloon sizes ranged from 5 to 10 mm with balloon-to-stenotic diameter ratios ranging from 2.6 to 4.2 (Table 1).

https://doi.org/10.1017/S1047951120002292 Published online by Cambridge University Press

Table 1. Patients' characteristics, diagnosis, and procedural details

	Diagnosis	Bilateral SVC[¶]	SPCA type (stenosis underlined)		Indication for Cath	Months from surgery	Weight (kg)	SVC diameter (mm)[†]	Adjacent PA diameter (mm)	Balloon size (mm)	SVC-PA anastomosis diameter (mm)			
			Right	Left							Pre	Post	Diameter on follow up Cath	Follow up duration (months)
1	HLHS (AH, MH); aortic arch hypoplasia; CoAo; perimembranous VSD	Yes	HF	BDG	Hypoxemia	2.4	7.8	4.1	3.4 (dist LPA)	5	1.3	2.6	2.8	2.3
2	TA and PA, hypoplastic pulmonary arteries	Yes	HF	BDG	Hypoxemia	2.2	7.7	4.1	6 (dist LPA)	5	1.2	3.4	5.5	11.9
3	HLHS (AA, MA)	Yes	HF	BDG	Pre-Fontan	23.3	12.1	8.8	5.4 (dist LPA)	10	4.2	6.2	6.8	3.1
4	DORV; subaortic VSD; moderate muscular VSDs; TV attachments to septal crest; PS	Yes	HF	BDG	Pre-Fontan	20.6	13.7	10.2	7.5 (dist LPA)	10	4.7	7.8	8.5	4.9
5	Heterotaxy; dextrocardia; RA isomerism; DORV; unbalanced CAVC with hypoplastic LV	Yes	BDG	BDG	BDG stenosis by Echo	2.8	9.3	5.2	4.8 (prox RPA)	7	2.2	4.7	6.6	27.7
6	Heterotaxy, TGA, unbalanced CAVC with hypoplastic RV; PAPVR	No	BDG	N/A	hypoxemia	3.4	9.2	8.1	5.3 (dist RPA) 4.6 (prox RPA)	6 + 6 [¥]	3.7	5.4	7.4	14.6
7	PA/IVS with severely hypoplastic RV, RV-dependent RCA	No	BDG	N/A	hypoxemia	1.3	7.7	6.8	4.2 (prox RPA)	7	3.3	4.4	8.3	29.3

[¶]No bridging veins in all cases.
†Measured cranial to the stenosis where diameter is considered to be normal and not affected by anastomosis.
[¥]Two-balloon technique was used in this patient.
AA, MA: aortic and mitral atresia; BDG: bidirectional Glenn; CAVC: common AV canal; CoAo: coarctation of aorta; DORV: double outlet right ventricle; HF: Hemi-Fontan; HLHS: hypoplastic left heart syndrome. AH, MH aortic and mitral hypoplasia; LV: left ventricle; PA: Pulmonary atresia; PA/IVS: pulmonary atresia intact ventricular septum; PAPVR: partial anomalous pulmonary venous return; PS: pulmonary stenosis; RA: right atrium; RCA: right coronary artery; SCPA: superior cavopulmonary anastomosis; SVC: superior vena cava. TA: Tricuspid atresia; TGA: transposition of great vessel; TV: tricuspid valve. VSD: ventricular septal defect.

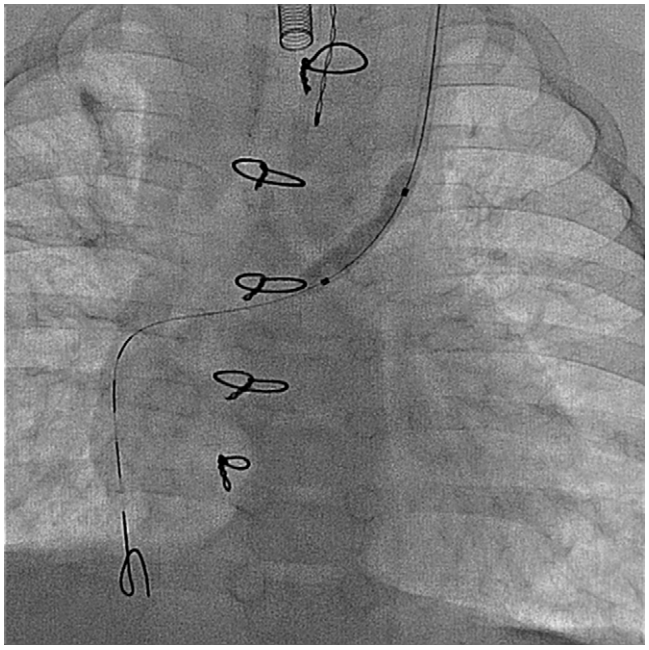

Figure 3. Patient #2, access to left BDG via left internal jugular vein.

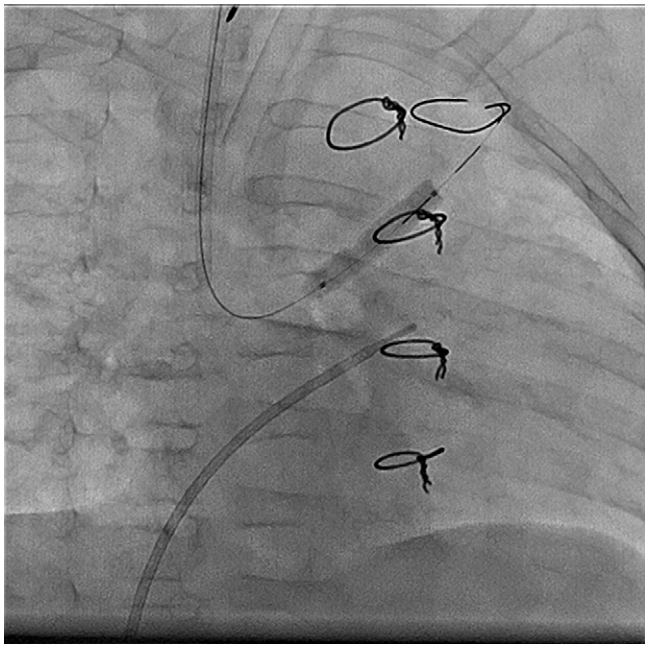

Figure 4. Patient #1, access to left BDG via right internal jugular vein.

Procedural results and follow-up

All procedures were successful, with no adverse events. Median stenosis diameter was 3.3 mm (range 1.2–4.7 mm) and increased to a median diameter of 4.7 mm (range 2.6–7.8 mm) after angioplasty. An example of a pre- and post-angioplasty angiogram is illustrated in Fig 1. All patients had a follow-up cardiac catheterisation with the follow-up interval ranging from 2 to 29 months. Indications for follow-up catheterisation were routine pre-Fontan assessments in four patients, hypoxia in one patient, recurrent pleural effusion in one patient, and evidence of recurrent stenosis by echocardiogram in one patient. First follow-up catheterisation was 2–27 months after the index procedure and showed a median diameter of 6.6 mm (range 2.8–8.5 mm). Measurements pre- and post-angioplasty and at subsequent follow-up cardiac catheterisations are illustrated in Fig 5. Only one patient had significant recurrence and required repeat balloon dilation (patient #1). Follow-up catheterisation and angiography showed sustained results in all other patients with further interval increase in the absolute diameter of the treated SVC/PA junction in all patients. When the SVC-PA junction diameter was indexed to body surface area (BSA), the interval growth was still evident, however, not proportional to the increase in BSA as it is typical in SCPA and Fontan patients. Therefore, we compared the ipsilateral branch PA diameter to the contralateral branch PA diameter supplied by the untreated non-stenotic SCPA in the patients with bilateral SVC pre-procedure and at follow-up cardiac catheterisation to assess if adequate growth was achieved. Fig 6 shows comparable interval size increase on both sides.

Discussion

SCPA is a common palliative procedure for complex cyanotic heart disease with univentricular anatomy and/or physiology with two surgical procedures to achieve SCPA: HF and BDG generally selected based on surgeon and/or centre preference. The HF operation includes augmentation of the PA[1,5] which may make SCPA stenosis less likely, as our experience of all SCPA stenosis occurring after BDG would suggest. Azygos vein ligation is not routinely performed as part of SCPA in our centre, which in the setting of SCPA stenosis may lead to significant hypoxemia from run-off into the inferior vena cava system. Echocardiography detected the SCPA stenosis in four of seven patients. The run-off through azygos vein and other collaterals may result in low-velocity non-turbulent flow making echocardiographic detection more difficult. The incidence of SCPA stenosis has not been specifically reported but may be included in reports of PA branch complications after SCPA. In our series, only 7 of 84 (8%) catheterised patients were found to have a significant stenosis that required angioplasty, all of them related to a BDG. Cnota et al. reported 2 out of 193 patients who underwent balloon angioplasty of SVC as an unanticipated intervention during hospitalisation for SCPA.[6] Reinhardt et al. reported SVC balloon angioplasty among other interventions after second-stage palliation.[4] It has been reported that in bilateral SVC, the caval veins are smaller, making the SCPA procedure technically more challenging[7] with different views on whether it affects outcomes.[8,9] In our series, five of seven patients had bilateral SVC and underwent bilateral SCPA. None of our patients underwent stent implantation in the SVC/PA junction because the trifurcating anatomy of the lesion and the small patient size preclude definitive stent angioplasty that can reach adult size for all three pathways without development of stenosis.

Interval growth was noted in the PA branch ipsilateral to the treated SCPA. It has been reported that PA growth is impaired following SCPA.[10] Therefore, in patients with bilateral SVC, we compared the PA branch diameter ipsilateral to the treated BDG to the contralateral PA supplied by the unobstructed SCPA (Fig 6) and found comparable growth.

Decompressing veno-venous collaterals occur in about 30% of patient after SCPA, with SVC and innominate vein being the most common sites.[11,12] Development of such collaterals correlates with

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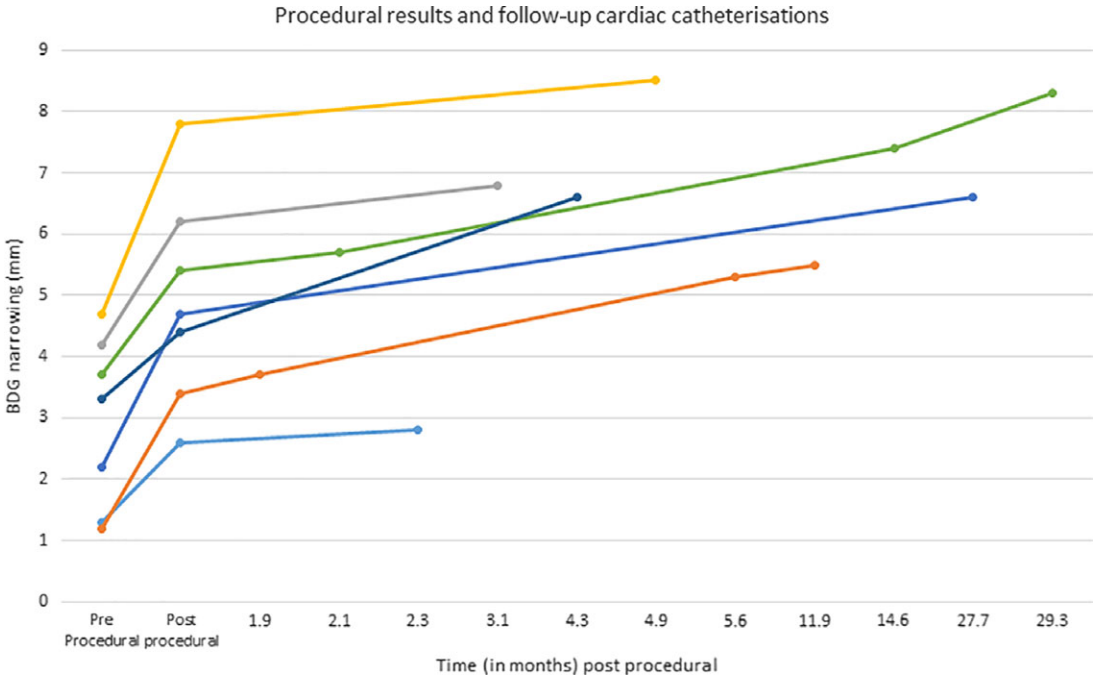

Figure 5. Diameter of the SVC-PA anastomosis by angiography before, immediately after, and at follow-up.

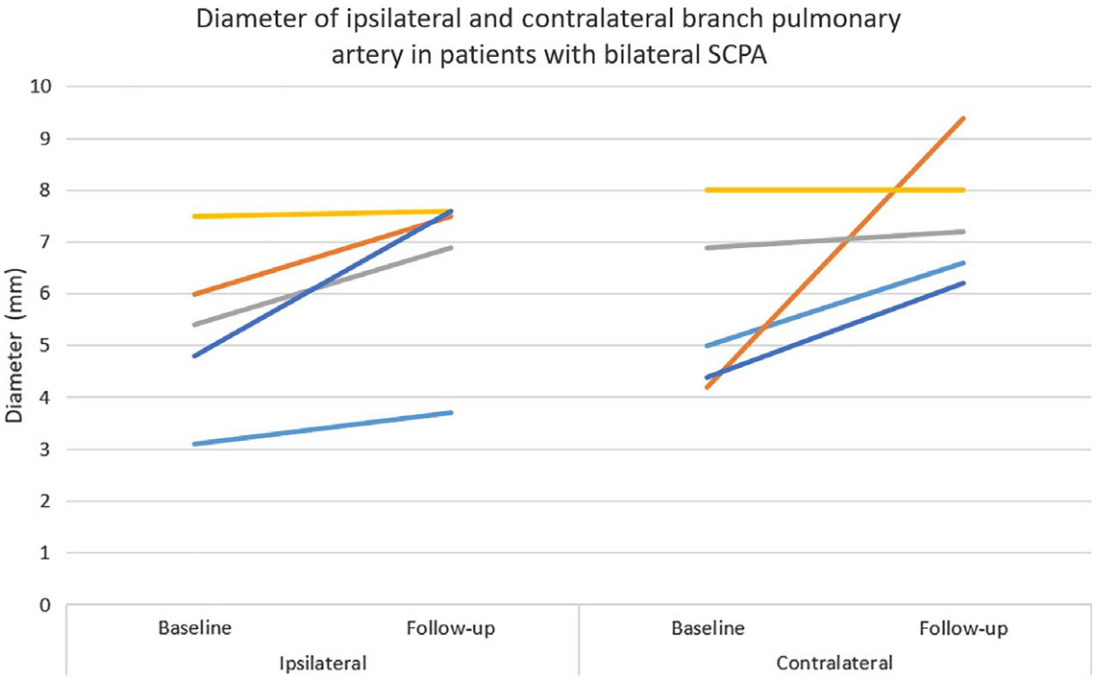

Figure 6. Diameters of ipsilateral and contralateral branch pulmonary artery (measured distal to SCPA and before the first branch) in the five patients with bilateral SVC before angioplasty and at follow-up.

the pressure gradient between SVC and right atrium.[11,12] It is likely that stenosis of the SVC-PA anastomosis can enhance decompressing collateral development, especially if the azygous vein is not ligated. Early treatment is therefore important clinically and our results suggest that balloon angioplasty can provide effective and lasting improvement. If the SCPA stenosis is contralateral to the Fontan side, our results suggest that balloon angioplasty can obviate the need for time-consuming remote patch augmentation at the time of Fontan operation, as we showed sustained results with further interval growth on follow-up in six of seven patients (Fig 5).

It has to be noted that our study is limited by its small sample size and its retrospective nature. A small number of patients who underwent SCPA returned to their referring institution and were

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not screened for potential stenosis. Although angiographic results are favourable, there are many confounders that precluded us from attributing clinical improvement exclusively to the procedure particularly in view of the small sample size.

Conclusion

Our small study suggests that transcatheter balloon angioplasty of the BDG anastomosis is feasible, safe, and may provide sustained improvement with only rare recurrence of stenosis and interval growth in most cases.

Financial Support. This research received no specific grant from any funding agency, commercial or not-for-profit sectors.

Conflicts of Interest. None.

Ethical standards. The study was approved by the institutional review board of the Alfred I duPont Hospital for Children.

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