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[Original article]

Percutaneous intervention for central shunts: new routes, new strategies

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Introduction In traditional locations, the standard Blalock-Taussig shunt presents numerous technical difficulties for percutaneous intervention. We changed our strategy to a central type shunt (Laks-type) with end-to-side pulmonary and side-to-side aortic anastomosis. The aim of this study was to determine whether this modified strategy would allow easier percutaneous manipulation in patients with small pulmonary arteries.

Methods All children with a stretchable central vascular graft who required any form of percutaneous intervention were prospectively enrolled in the study.

Results Eleven infants were evaluated a median time of 3 months (range 0.9-4.4) following initial shunt placement; the median weight at intervention was 5.7 kg (range: 4.0 – 10.0). All shunts (100%) were easily and swiftly entered without the need for special catheters or co-axial systems. In four patients other interventions in distal pulmonary arteries were first performed: cutting balloon treatment in three and balloon angioplasty of peripheral pulmonary artery stenosis in one. The shunts were then augmented with a stent with a diameter increasing from 3.5 ± 0.4 mm to 4.7 ± 0.8 mm and saturation increasing from 76% (range: 69-88) to 84% (range: 77-88) ($P < 0.05$). Several months later, two children required further interventions that could easily be performed via the stented shunts. No complications were observed.

Conclusions The Laks-type shunt provides easy access for percutaneous procedures of the distal pulmonary arteries including cutting balloons; this shunt can predictably be expanded to augment pulmonary flow. This study highlights how co-operation between the interventionalist and the surgeon can improve strategies to manage these difficult patients.

Keywords *Hypoplasia pulmonary artery – intervention – shunt – stent – angioplasty.*

INTRODUCTION

In the current era, primary repair in the neonatal period has become the procedure of choice for a considerable number of congenital cardiac defects. For complex lesions requiring catch-up growth by augmentation of blood flow to the lungs, the most common palliative procedure still remains a systemic-to-pulmonary artery shunt. Based on experience with univentricular hearts, the majority of cardiothoracic surgeons would nowadays prefer sternotomy with central shunt placement in patients with small or diminutive pulmonary arteries. Smaller diameter shunts are generally chosen in order to preserve ventricular function and to protect it from volume overload[1]. However, it has also become clear that the size of the pulmonary vascular bed is an important determinant for a good Fontan circulation and strategies should aim at maximizing growth of the fragile pulmonary vascular bed[2].

Most shunts allow little or no compensation to alter flow in parallel with growth. It would be beneficial for infants to improve flow in these small shunts as it would allow development of the important pulmonary vascular

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bed as well as somatic growth of the child, simplifying subsequent surgery.

Numerous complications such as thrombosis, kinking and distortion of the vessels have been described as a result of systemic to pulmonary artery shunts[3]. Several publications have shown that balloon dilation and stent implantation can be used to treat stenosed shunts[4-6]. We have demonstrated that stretchable Gore-Tex® vascular grafts (W.L. Gore & Associates, Inc. Flagstaff, Arizona, US) can be safely and effectively enlarged to improve blood flow through a shunt[7]. However, a number of technical difficulties arise when a modified Blalock-Taussig shunt originating from either the subclavian or brachiocephalic artery needs percutaneous treatment. These shunts are often difficult to enter due to the acute angles at the proximal anastomosis, especially with cutting balloons; also, when stent implantation is attempted, stents may get stuck or become dislodged. Large bore delivery catheters are also frequently required to facilitate stent delivery. Since these shunts frequently require augmentation at the arterial end, stents tend to protrude into the lumen of the vessels after delivery, making re-entry into the shunt and/or additional procedures in the future almost impossible.

As a result, we adapted our overall strategy. We decided to aim for smaller dilatable shunts that can easily be accessed percutaneously allowing multiple subsequent re-interventions if required. Since sternotomy is currently preferred, we opted to use stretch Gore-Tex® vascular grafts anastomosed end-to-side to the pulmonary artery and side-to-side to the anterior aorta in the fashion as described by Laks in patients requiring shunt palliation[8,9] (figure 1). The aortic anastomosis is typically large, thereby obviating the need for stenting the orifice.

The aim of this study was to determine whether this modified strategy would allow easy access and percutaneous manipulations in patients with small pulmonary arteries.

METHODS

Patients

All children with a stretchable central vascular graft requiring any form of percutaneous intervention, were prospectively entered into the database. Patient records were used to obtain catheterization and follow-up data. Digital measurements were performed using an IMPAX viewer (Agfa Heartlab®). The study was conducted in accordance with the local ethics committee guidelines and fully informed consent was obtained. Pulmonary artery diameters were not routinely measured during follow-up since this would have required additional catheterizations and exposure to radiation in these small infants.

Technique (figures 1-4)

The central shunt, consisting of thin-walled stretch Gore-Tex® vascular grafts (W.L. Gore & Associates, Inc.Flagstaff, Arizona, US) which are made of polytetrafluoroethylene (PTFE), was connected end-to-side

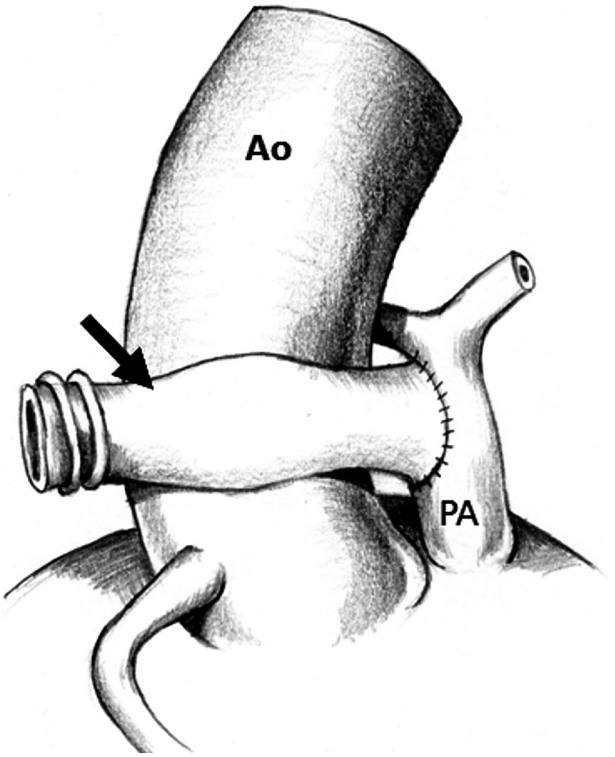

Fig. 1 Diagram of Laks-type shunt
Ao (aorta); PA (main pulmonary artery). Arrow indicates Goretex side-by-side shunt with surgical clips.

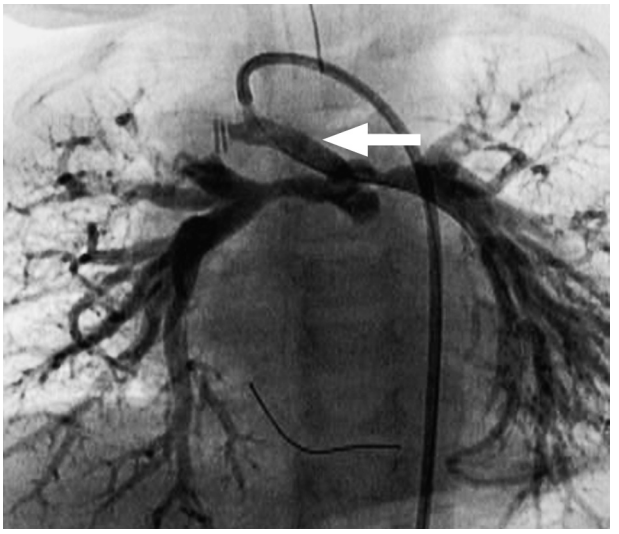

Fig. 2 Angiogram of shunt (n° 10)
Frontal projection demonstrating Laks-type shunt (arrow) before intervention. This patient required cutting balloon treatment of both pulmonary arteries.

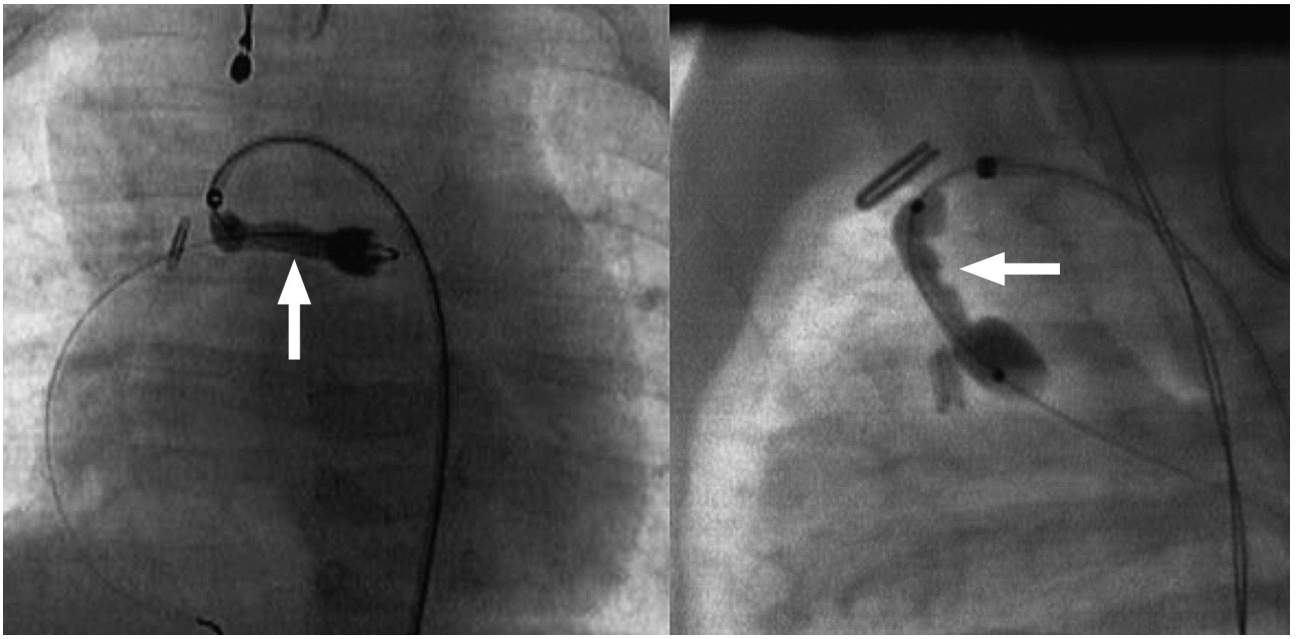

Fig. 3 Balloon interrogation of shunt (n° 9 and 10)
Frontal (left) and lateral (right) projection. Tyshak balloon inflated in central shunt, note clear delineation of length and widely patent proximal and distal orifices. The angle of the 4-F sheath illustrates the favourable angle of access. Arrow demonstrates outline of shunt.

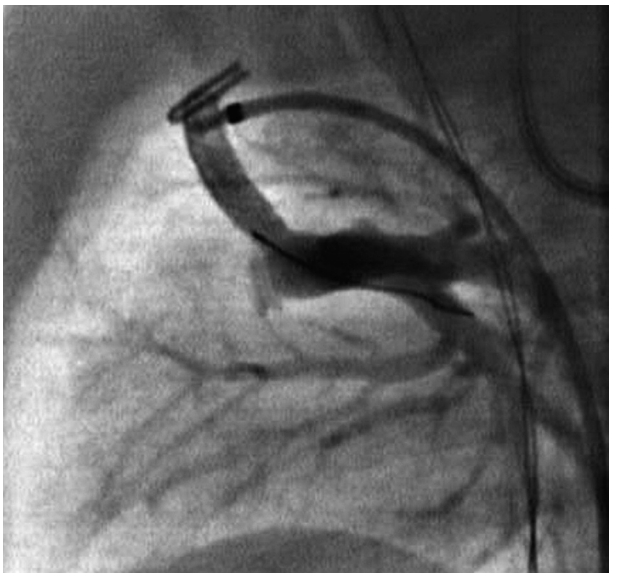

Fig. 4 Successful stent implantation (n° 9)
Angiography shows successful stent implanted and enlargement of shunt diameter.

to the pulmonary artery and side-to-side to the anterior aspect of the aorta at a height that would allow easy passage for a catheter. At least two surgical clips were placed to seal the shunt.

Procedures were performed under general anaesthesia. The femoral arteries were punctured and standard 4-F short sheaths inserted. The shunt was then entered, usually by means of a 4-F vertebralis type catheter. We typically used a microcatheter with soft guide (Progreat™, Terumo, Europe N.V., Belgium) to select a distal pulmonary artery branch and a stiff guide wire (0.014") was then anchored in this vessel. The sheath was at that point exchanged for a 4-F 45-cm long Cook sheath (Cook Medical, Bloomington, US). The required procedures in distal pulmonary arteries were then performed. The shunt was enlarged; it was first interrogated using a mildly oversized compliant balloon (typical 6-mm diameter, 20-mm long Tyshak, NuMED, Hopkinton, NY, USA). This is an important step in the procedure not only because it demonstrates the underlying anatomical substrate, but also allows determination of the exact length of the required stent. Stents were delivered and a pressure insufflator used to ensure the correct diameter (figures 1-3); a 3.5-mm shunt can be augmented to 4.9 mm, a 4.0 mm shunt up to 5.5 mm[7]. Antibiotics and heparin were administered according to routine protocol. Aspirin (1-2 mg/kg/day) was given at discharge.

Statistical analysis

Data was captured using Excel spreadsheets and statistical analysis was performed using standard statistical software (SPSS for windows, SPSS Inc., IBM Company, Chicago, Illinois, US, version 18). A student-*t* test was

used to compare normally distributed data. Continuous data were expressed as medians with the minimum and maximum values. A *P*-value <0.05 was considered significant.

RESULTS

Table 1 shows patient and procedural characteristics. Over a period of four years 19 central shunts were inserted; in 11 patients flow augmentation was required during follow-up. This subgroup forms the basis of this report. The Laks shunt was performed at a median age of 1.3 months (range: 0.2-16.0) with a median weight of 3.6 kg (range: 1.6-8.9). The majority of infants received 3.5-mm shunts and was diagnosed with variations of univentricular heart disease or tetralogy of Fallot with pulmonary atresia-type lesions.

Flow augmentation by percutaneous interventions was required a median time of 3 months (range: 0.9-4.4) after original shunt placement; median age 4.6 months (range: 2.0-20.0) , weight at intervention 5.7 kg (range: 4.0-10.1). The predominant indications for intervention were arterial desaturation, persistent peripheral pulmonary artery stenosis and hypoplasia increasing the risk or excluding the possibility of corrective surgery.

All shunts (100%) were easily and swiftly entered without the need for special catheters or co-axial systems – the angle of attack facilitated shunt entry and balloon positioning. In four patients other interventions were first performed: cutting balloon treatment (patients 4, 6 and 10) of 3.5 mm and balloon angioplasty (patient 9) of peripheral pulmonary artery stenosis. All balloons could easily enter the shunt and the branch pulmonary arteries.

Shunt augmentation by placement of a stent was easy and predictable; no patient required a second stent. No stent got stuck upon entry of the shunt or was dislodged. Coronary artery stents were used and were mostly 1.0-1.5 mm larger than the original shunt diameter. Length of the stents (as determined by interrogation balloon) varied from 12-28 mm (table 1). Five stents were post-dilated with a larger balloon at high pressure (patients 1, 5, 8, 9 and 10). The shunt diameter increased from 3.5 ± 0.4 mm to 4.7 ± 0.8 mm; the final shunt diameter could on average be increased by 1.2 ± 0.6 mm ($P < 0.001$). Percutaneous oxygen saturations improved significantly from a pre-procedural median of 76% (range: 69-88) to 84% (range: 77-88) ($P < 0.01$) (figure 4). All infants remained stable throughout the procedure; all femoral arteries were patent on follow-up.

Table 1 Patient and procedural characteristics and outcome

n°	Diagnosis	Surgical shunt				Stent placement					
		Diameter (mm)	CPB	*Weight (kg)	*Age (mo)	Int (mm)	Diam(mm)	Length (mm)	Type	Outcome	Time (mo)
1	PS, RV hypopl, TGV, UVH	3.0	n	4.0	2.0	6	4.0	12	Integrity (Medtronic)	BDG	3.6
2	DORV, PS, TiV abnormality	3.0	y	4.9	4.6	6	5.0	16	Liberte (Boston)	repair	7.6
3	UVH TA	3.5	y	5.7	4.6	6	5.0	18	Prokinetic Energy (BIOTRONIK)	BDG	8.9
4	PA-VSD	4.0	n	8.9	11.1	6	5.0	24	Liberte (Boston)	repair	8.4
5	UVH MS, SHONE	4.0	y	8.0	20.1	7	5.8	24	Liberte (Boston)	BDG	10.7
6	PA-VSD	4.0	n	6.2	4.4	6	5.0	20	Liberte (Boston)	repair	11.2
7	DORV TGA PA	3.5	n	4.7	2.9	6	5.6	18	Prokinetic Energy (BIOTRONIK)	BDG	7.9
8	PA-VSD	3.5	n	10.0	14.2	5	4.0	28	Omega (Boston)	palliation	12.3
9	DILV, DORV, UVH	3.5	n	5.0	5.2	5	4.5	16	Omega (Boston)	BDG	5.3
10	PA VSD	3.5	n	7.4	10.5	7	4.0	24	Liberte (Boston)	repair	17.9
11	PA, L-TGV, UVH	3.5	n	5.0	2.1	6	4.5	20	Liberte (Boston)	BDG	2.3

Abbreviations: PS: pulmonary stenosis, RV: right ventricle, TGV: transposition of great vessels, UVH: univentricular heart, DORV: double outlet right ventricle, TiV: tricuspid valve, TA: tricuspid atresia, PA–VSD: pulmonary atresia with ventricular septal defect, MS: mitral stenosis, DILV: double inlet left ventricle, FU: follow-up, BDG: bidirectional Glenn, int: interrogation balloon.
*: weight and age at stent placement, n: no, y: yes (note 1 patient only normothermic bypass), diam: diameter stent.

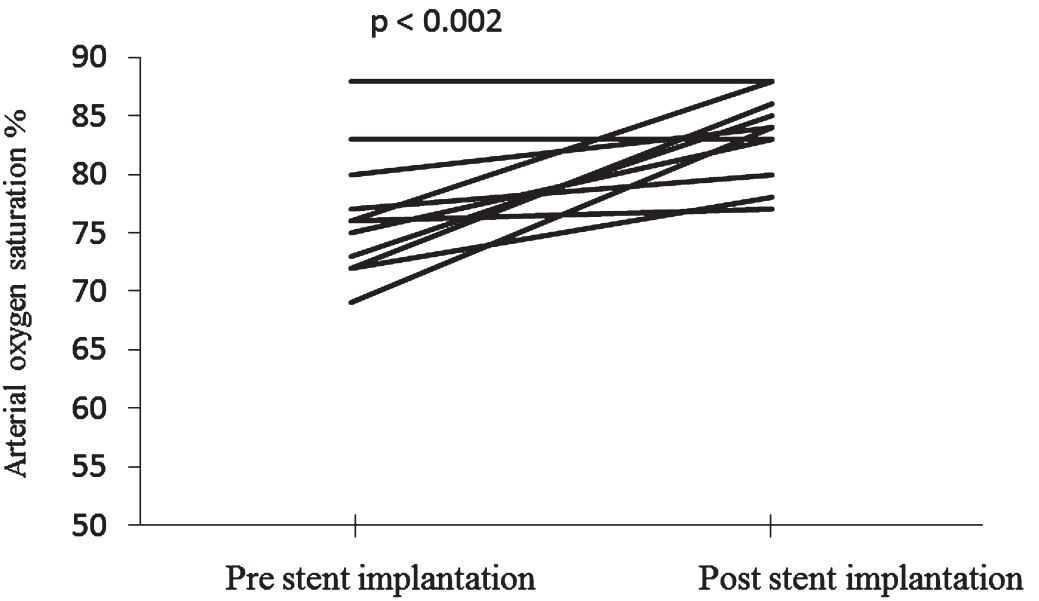

Fig. 5 Improvement in oxygen saturations Arterial oxygen saturations before (pre) and after (post) stent implantation.

FOLLOW-UP

In two patients (n° 4 and 10) two further procedures in the distal vascular bed were performed at a later session following stent implantation. The stented shunt was easily entered and no problems were encountered during the procedures.

All patients proceeded to either palliative or reparative surgery. The median saturation pre-operatively was 82% vs 84% after shunt intervention ($P=0.82$). Surgery was delayed a median of 12.3 months (range: 5.3-26.9) ± 4.9 (table 1). No deaths or episodes of stent thrombosis were reported during the follow-up period.

DISCUSSION

The results of this study show that our newly adopted strategy is feasible. All central shunts could easily be accessed for various transluminal procedures on the pulmonary arteries. None of the shunts in the series showed any signs of stenosis at the two anastomotic sites. Stent augmentation of the shunt was easy and predictable. Saturations improved significantly following stenting and the lifespan of the shunts could be extended with several months.

In patients with a high need for pulmonary artery rehabilitation or in children with very small pulmonary arteries, the Laks-type central shunt is now our preferred procedure. This is in agreement with the findings of other groups using a side-by-side shunt who demonstrated good patency as well as pulmonary artery growth[8,9]. If patients do well, they proceed to the next phase of surgery and if additional flow is required, percutaneous interventions can easily be performed. It allows effortless access to both the left and right pulmonary arteries as well as easy access for stent enlargement of the shunt with less distortion of the branch pulmonary arteries. Access to the shunt and pulmonary arteries was much easier compared to the previous strategy of modified Blalock-Taussig shunts – there were no acute angles to be crossed and stents could straightforwardly be advanced into position. No stent was dislodged or required additional support with delivery catheters or sheaths. As a result, smaller 4-F arterial access systems only could be used compared to our previous strategy where 6-F systems were often required to traverse the acute angles[7]. This of course reduces the risk of damage to the small femoral arteries in these children. The complex nature of this group of patients is emphasized by the fact that cutting balloons were required in almost a third of children for treatment of peripheral pulmonary artery stenosis. It is noteworthy that these bulky and difficult to manipulate rigid balloons could be passed through the shunt into the desired peripheral vessel without much difficulty.

Stenting the central shunt in our group of patients clearly prolonged shunt lifespan: 5 patients proceeded to either reparative surgery or univentricular palliation and in the others surgery has been delayed for a couple of months with little change in percutaneous saturation. The effectiveness of the procedure is confirmed by the significant improvement in percutaneous oxygen saturations and supports our previous findings that stretchable shunts can be enlarged to improve flow[7].

The ideal palliative shunt would regulate flow according to somatic growth, but that is not possible with fixed diameter shunts. Central shunts are nowadays preferred in patients with diminutive pulmonary arteries since it allows a more homogeneous distribution of blood flow to the lungs and it makes the anastomosis to the small pulmonary artery easier[8-12]. Complications of systemic to pulmonary artery shunts occur both early and late with an overall mortality in the region of 15%. Shunt dysfunction, comprising several causes including thrombosis, distortion and early flow restriction have been described in up to 42% of newborns undergoing shunting[13,14]. It is quite likely that these adverse events would occur even more frequently in low-birth-weight infants with hypoplastic pulmonary arteries. Shunt-related complications have been shown to be more common in shunts of less than 4 mm [15,16].

Patients also included heterotaxy syndromes and complex univentricular heart abnormalities which have been identified as specific risk groups and it is therefore not unusual to expect shunt-related problems in these patients over time[14]. Particularly in these children it would be beneficial to extend shunt lifespan and treat underlying pulmonary arterial problems, since it would theoretically allow improved pulmonary arterial and somatic growth, improving the substrate for a univentricular repair. Our findings correspond favourably to the experience of this type of side-to-side aortopulmonary shunts by Barozzi et al., who demonstrated harmonious growth of the pulmonary arteries using the Nakata index[17].

TECHNICAL CONSIDERATIONS

Although access was easy, we found the use of a microcatheter inserted via a 4-F angled catheter (e.g. vertebralis) extremely helpful to assist in the acquisition of stable guide wire position in a distal pulmonary artery branch[18]. The use in small infants of a 45-cm 4-F sheath was also extremely valuable – apart from the limiting vascular access to a small diameter, it served to stabilize the system, angiograms could be performed via the side port and it is large enough to allow delivery of small premounted coronary artery stents. Procedures of the pulmonary artery and branches should be carried out before stent enlargement is performed in order to prevent dislodgement of the stent.

The shunt flow in the standard modified Blalock-Taussig shunt is controlled by the orifice of the proximal artery, the sites of anastomosis, shunt diameter and length; in contrast, flow in the Laks-type shunt, due to the nature of the anastomoses, is controlled by diameter and length. The proximal aortic anastomosis is typically non-restrictive while obviating the need to stent the anastomosis itself and averting protrusion of the stent into the aorta. Interrogation with a mildly oversized semi-compliant balloon allowed to accurately determine the required stent length. It is therefore not surprising that only one stent was necessary to enlarge the cross-sectional area of a shunt. Based on the results of our previous study which included bench-testing, we generally selected stents to be about 1-1.5 mm larger than the original shunt[7]. Although no complications occurred, it should be noted that dissection and aneurysm formation have been described.

LIMITATIONS

This study suffers from the limited numbers and cross-sectional nature of its design. Pulmonary artery growth was not measured since it was not an objective of this study. Yet, it was considered but not performed because the investigators wanted to limit radiation, bearing in mind that multiple future procedures were expected. However, as pointed out, Barozzi demonstrated a satisfactory growth of the pulmonary arteries in their study[9]. Patients were not randomized since the aim was to limit the need for repeated surgical procedures.

CONCLUSIONS

Percutaneous intervention in stretchable central shunts (Laks-type) in patients with small pulmonary arteries is feasible and safe. The Laks-type shunt provides easy access for percutaneous procedures to the branch pulmonary arteries. Pulmonary arteries can be rehabilitated, blood flow to the lungs and arterial oxygen saturations can significantly be improved and the lifespan of the original shunt prolonged. This study highlights how co-operation between the interventionalist and the surgeon can improve strategies to manage these difficult cases and reduce shunt-related morbidity.

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CONFLICT OF INTEREST

Marc Gewillig is proctor for Numed and Medtronic.

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