Transcatheter palliation of congenital heart disease with reduced pulmonary blood flow

Giuseppe Santoro, Maurizio Cappelli Bigazzi, Giuseppe Caianiello*, Marianna Carrozza, Maria Teresa Palladino, Gaetano Palma*, Maria Giovanna Russo, Carlo Vosa*, Raffaele Calabrò

*Division of Cardiology, *Division of Pediatric Cardiac Surgery, Second University of Naples, Naples, Italy*

Key words:
Congenital heart disease; Patent ductus arteriosus; Shunts; Stents.

Background. **Although surgical shunt is still considered as the best palliation of congenital heart disease with reduced pulmonary blood flow, stent implantation may be technically simpler, safer and more cost-effective than surgery in high-risk patients. This study evaluated the feasibility and results of this option in patients with duct-dependent pulmonary blood flow or systemic-to-pulmonary shunt malfunction.**

Methods. **Between April 2003 and July 2004, 9 patients (age 11 days-52 years, weight 2.1-52 kg) with complex congenital heart disease underwent stent implantation inside the patent ductus arteriosus (4 patients) or a stenotic surgical shunt (5 patients).**

Results. **The stenting procedure was successfully completed in all cases. The procedural time was 162 ± 36 min (range 90-225 min). The fluoroscopy time was 33.8 ± 6.8 min. No patient died. The morbidity rate was 22.2% (1 patient had local infection at the site of puncture and 1 had transient femoral artery pulse loss). After the procedure, the ductus/shunt diameter increased from 1.2 ± 0.6 to 3.6 ± 0.6 mm ($p < 0.0001$) and oxygen saturation improved from 74.0 ± 6.5 to 85.2 ± 3.3% ($p < 0.01$). Three patients underwent corrective surgery without technical problems after 8.0 ± 1.0 months while oxygen saturation remained constantly > 80% in patients still waiting for surgical repair (follow-up 5.3 ± 3.1 months).**

Conclusions. **Stent implantation is a technically feasible, safe and effective palliative option in high-risk surgical patients with congenital heart disease and reduced pulmonary blood flow. Although larger series are required to define the cost-effective clinical impact of this therapeutic option, it is reasonable to hypothesize a further extension of its indication even to elective and low-risk surgical patients.**

(Ital Heart J 2005; 6 (1): 35-40)

Received September 22, 2004; revision received November 17, 2004; accepted December 7, 2004.

Address:
Dr. Giuseppe Santoro
Via Vito Lembo, 14
84131 Salerno
E-mail:
santoropino@tin.it

Introduction

Despite current trends toward early primary repair, surgical shunt is still considered as an invaluable palliative option for cyanotic congenital heart malformations in high-risk patients[1,2], as well as an important component of complex staged surgical approaches[3]. However, pleural effusion, chylothorax, phrenic and vagal nerve palsy and distortion and differential growth of the pulmonary arteries are well-known shunt-related complications potentially increasing the morbidity and mortality of corrective surgery particularly in small neonates and infants[1,2,4,5]. Over time, technical improvements have rendered the transcatheter approach a reliable alternative to surgery, either as initial palliation of duct-dependent cardiac malformations or as a rescue option for surgical shunt malfunction[6-20]. This report describes the experience from a tertiary referral center in terms of the feasibility and results of percutaneous palliation by stent implantation of congenital heart malformations with pulmonary hypoperfusion.

Methods

Patient population. Between April 2003 and July 2004, 9 patients with duct-dependent pulmonary blood flow (n = 4) (Fig. 1) or surgical shunt malfunction (n = 5) (Figs. 2 and 3) underwent percutaneous palliation by patent ductus arteriosus (PDA)/shunt stenting. In all cases, the patients or parents gave their informed consent to the procedure. The patients' clinical and procedural data are summarized in table I. Indications to interventional catheterization were a high-risk profile (n = 6), an elective offer as alternative to surgery (n = 2), and the refusal of any surgical approach (n = 1). Patients were considered as having a high-

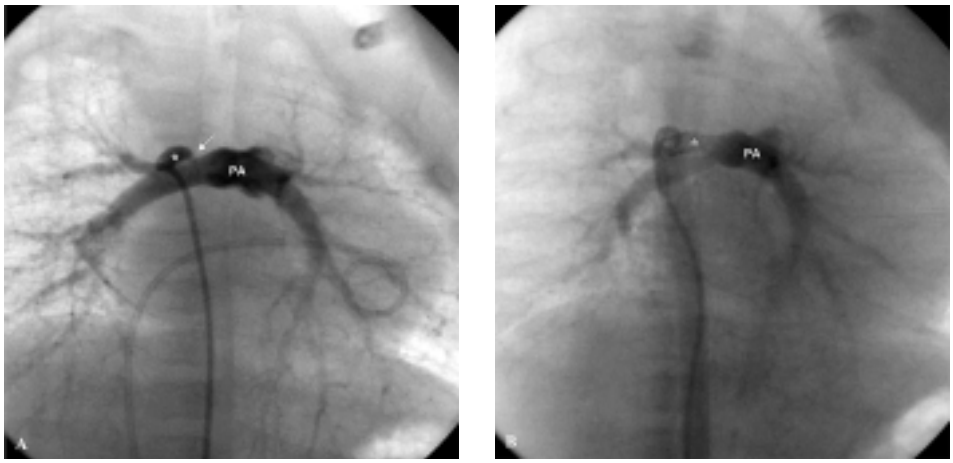

Figure 1. *Stent implantation within the patent ductus arteriosus in a patient with univentricular heart and severe pulmonary subvalvular and valvular stenosis.* A: *the patent ductus arteriosus* (*) *has been imaged from the aortic side, outlining a severe stenosis at its pulmonary end* (arrow). B: *the ductal stenosis has been completely relieved by stent implantation. PA = pulmonary artery.*

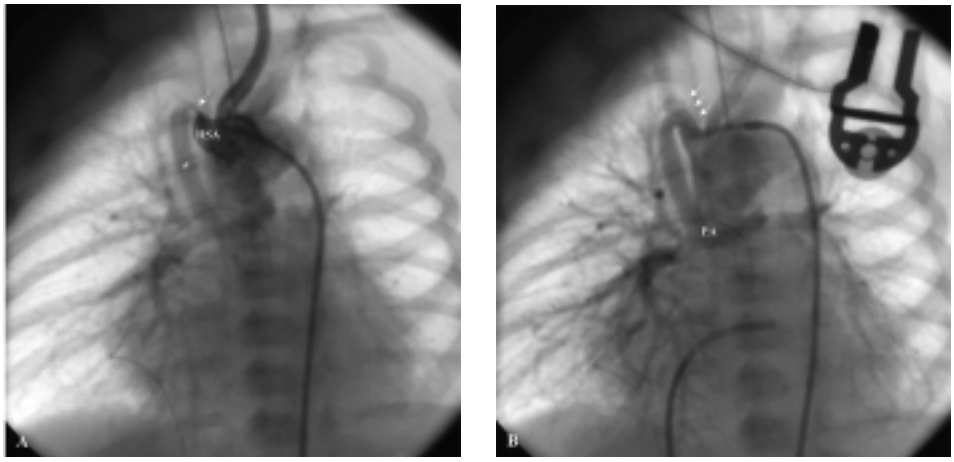

Figure 2. *Stenting procedure to relieve a tight stenosis of a Blalock-Taussig shunt in a patient with hypoplastic left heart syndrome submitted to the Norwood stage I procedure.* A: *the obstruction* (arrow) *has been imaged at the level of the subclavian artery and does not involve the proximal anastomosis of the modified Blalock-Taussig shunt* (*). B: *since the patient had been operated just a few days before, the stent was implanted inside the right subclavian artery (RSA)* (arrows)*, with complete relief of the stenosis. PA = pulmonary artery.*

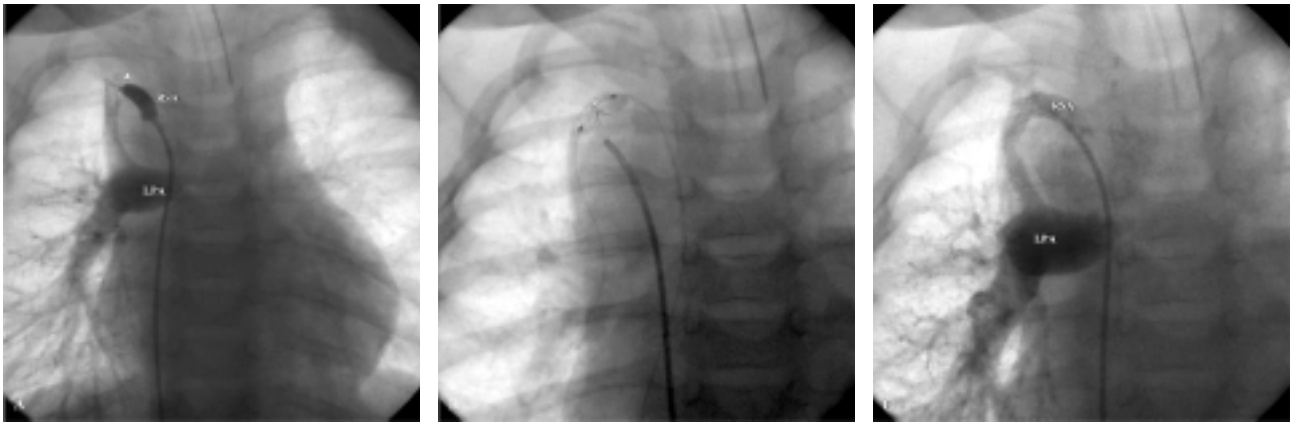

Figure 3. *Stent implantation to treat pulmonary hypoperfusion in a patient with hypoplastic left heart syndrome submitted to the Norwood stage I procedure. This patient had been operated a few weeks before. For this reason, we decided to deploy the stent at the right subclavian artery (RSA)/Blalock-Taussig shunt junction.* A: *the obstruction has been clearly imaged at the proximal Blalock-Taussig shunt anastomosis* (*). B: *a 3.5 mm long coronary stent has been deployed across the junction* (*)*, with complete resolution of the stenosis* (C). *LPA = left pulmonary artery.*

Table I. Clinical and procedural data at stent implantation.

Patient	Diagnosis	Previous treatment	Age	Weight (kg)	Stenting site	Stent (length × diameter) (mm)	Final stent diameter (mm)	Follow-up (months)	Corrective surgery
#1	ToF with pulmonary artery discontinuity due to LPA origin from the PDA	None	16 days	2.1	PDA (tubular shape)	3.0 × 12	4.2*	7	Yes
#2	HLHS	Norwood stage I	11 days	3.2	Right subclavian artery	3.5 × 8	3.5	9	BCPA
#3	UVH with mitral stenosis and pulmonary stenosis	Rashkind atrioseptectomy	84 days	3.9	PDA (tubular shape)	3.5 × 12 3.5 × 8	3.6	8	BCPA
#4	ToF	Classic left BT shunt	52 years	52	Left BT shunt	4.5 × 18	4.5	5	No
#5	PAIVS with aortic valve stenosis	RF pulmonary valvotomy	12 days	2.9	PDA (tubular shape)	3.5 × 13	3.5	5	No
#6	Situs inversus. cTGA with VSD and PA. Lusory right subclavian artery	Right and left modified BT shunts	17 months	12	Right subclavian artery/ BT shunt junction	4.5 × 8 3.5 × 8	3.5	9	No
#7	HLHS	Norwood stage I	29 days	3.5	Right subclavian artery/ BT shunt junction	3.5 × 8	3.5	9	No
#8	PAVSD	Right BT shunt	7 months	6	Right BT shunt	3.5 × 12 3.5 × 12	3.9	2	No
#9	PAIVS	RF pulmonary valvotomy	27 days	3.3	PDA (tubular shape)	2.5 × 8	2.2	2	No

BCPA = bidirectional cavo-pulmonary anastomosis; BT = Blalock-Taussig; cTGA = corrected transposition of the great arteries; HLHS = hypoplastic left heart syndrome; LPA = left pulmonary artery; PA = pulmonary atresia; PAIVS = pulmonary atresia with intact ventricular septum; PAVSD = pulmonary atresia with ventricular septal defect; PDA = patent ductus arteriosus; RF = radiofrequency; ToF = tetralogy of Fallot; UVH = univentricular heart; VSD = ventricular septal defect. * after stent re-dilation.

risk profile if they had a low weight (patient #1), a history of critical conditions early after demanding surgery (patients #2 and #7), an unusual anatomic arrangement (patient #5), and previous repeat palliative surgical approaches (patients #6 and #8). Patients #3 and #9 underwent elective PDA stenting in alternative to surgery following thorough parental counseling. The adult patient (#4) had been submitted to a classic Blalock-Taussig shunt procedure at the age of 9 years and refused any surgical option. Our policy is to submit patients with pulmonary valve atresia and intact ventricular septum to elective PDA stenting about 2 weeks after an initial radiofrequency valve perforation performed in case of failure to wean from prostaglandin infusion. After stent implantation, the patient was submitted to clinical and echocardiographic evaluation on a monthly basis and cardiac catheterization was planned just before corrective surgery.

Interventional procedure. Interventional catheterization was performed under general anesthesia in all but 1 patient. In case of a duct-dependent pulmonary circulation, prostaglandin infusion was stopped 4-6 hours before the procedure in order to achieve a stable duct constriction and hence enable a good grip of the stent after deployment. No patient presented with pre-procedural hypoxemia consequent to duct constriction during prostaglandin withdrawal or during stent positioning inside the PDA. The PDA or surgical shunt morphology, size and length were assessed in multiple views, accounting for the angiographic magnification. Stent implantation was performed using a femoral artery access in 8 patients and a right carotid artery access through a cut-down entry in a low-weight patient (#1) with the PDA originating from the inferior surface of the aortic arch. After PDA/shunt visualization, a 0.014" coronary guide-wire (ATW, Cordis Corporation, Miami, FL, USA) was negotiated through the stenotic vessel/conduit and anchored in a distal pulmonary artery branch. Stent positioning and deployment were angiographically-guided through a 23 cm-long 4F sheath (Avanti-plus Introducer, Cordis Corporation, Miami, FL, USA), so avoiding the use of larger guiding catheters. In case of duct stenting, the stent length was selected to cover the entire length of the PDA, while the stent diameter was individually chosen on the basis of the patient size, the pathophysiologic picture and the expected time of palliation. However, in our series, the stent diameter was always < 75% of the indicated surgical shunt size. When a surgical shunt stenting procedure was performed, the selected stent length and diameter were such as just to cover the stenosis and restore the stenotic segment to the original nominal size. The shunt stenosis was located on the subclavian artery side in patients #2, #6, and #7, in the middle of the polytetrafluoroethylene conduit in patient #8 and on the pulmonary artery side in patient #4. In patient #2, the stenosis did not involve the shunt origin and was reverted by stent implantation inside the subclavian artery (Fig. 2). In patients #6 and #7, the stenosis was located at the subclavian artery/shunt junction and was treated by an overriding stenting. In patient #8, two stents were deployed inside the prosthetic conduit, while in patient #4 the stent was implanted at the left subclavian artery/left pulmonary artery junction. Regardless of the site of implantation, no technical difficulties were encountered during the stenting procedure. After stent deployment, multiple view angiographic images were obtained to ensure that the stent had been correctly placed. Heparin infusion was continued for 24 hours at the dose of 100 IU/kg/day, followed by aspirin treatment at 3-5 mg/kg/day for 6 months.

Results

The stenting procedure was successfully completed in all patients. Twelve coronary stents were implanted in 9 patients (one stent in 6 patients and two stents in 3 patients). The procedural time was 162 ± 36 min (range 90-225 min). The fluoroscopy time was 33.8 ± 6.8 min. No patient died. The morbidity rate was 22.2% (1 patient developed a local infection at the site of puncture and 1 patient had transient femoral artery pulse loss). Stent implantation increased the PDA/shunt diameter from 1.2 ± 0.6 to 3.6 ± 0.6 mm ($p < 0.0001$) and improved percutaneous oxygen saturation from 74.0 ± 6.5 to 85.2 ± 3.3% ($p < 0.01$). After the procedure, 1 patient (#5) had to be prescribed anticongestive therapy for a few weeks, due to a generous post-procedure duct flow, while the remaining patients showed an adequate, balanced pulmonary blood flow. With regard to the mid-term follow-up outcome, 1 patient (#1) was submitted to stent re-dilation 4 months following the initial stent implantation, in order to fit the duct size to the pulmonary artery growth. Overall, pre-surgical catheterization was performed in 4 patients, showing an increase of the Nakata index from 233 ± 110 to 336 ± 92 mm/m^2 (p = NS). Three patients underwent corrective surgery, without any technical problem, after 8.0 ± 1.0 months. At surgery, the stent was completely removed in one patient (#1), with a patch reconstruction of its aortic insertion and a termino-lateral anastomosis of the left pulmonary artery on the main pulmonary artery. In the other 2 patients (#2 and #3) the stent was left in place: in the first one due to its location inside the right subclavian artery, in the second one after clipping and suturing of the ductus arteriosus. The patients still awaiting corrective surgery (#4, #6, #7, and #8) or being followed up in view of a spontaneous improvement (#5 and #9) had a percutaneous oxygen saturation constantly > 80% (Fig. 4), after a mean follow-up of 5.3 ± 3.1 months (range 2-9 months).

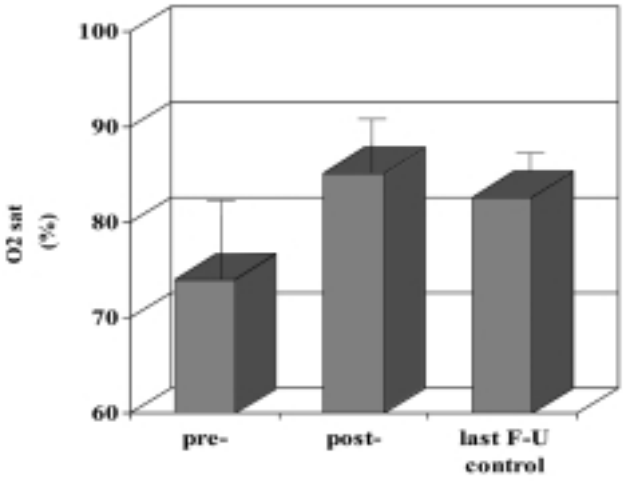

Figure 4. *Early and mid-term results of stent implantation within the patent ductus arteriosus/surgical shunt in terms of percutaneous oxygen saturation (O_2 sat). F-U = follow-up.*

Discussion

Post-natal ductus arteriosus patency is essential in congenital heart disease with a pulmonary or systemic duct-dependent circulation. In this setting, surgical shunting is still considered as the cornerstone palliative approach to pulmonary hypoperfusion[1,2], as well as an invaluable part of staged approaches to some complex congenital heart malformations[3]. Over time, the idea of using the ductus arteriosus as a natural conduit instead of a surgical shunt was initially put into practice by formalin infiltration[15] and later by transcatheter techniques, such as balloon angioplasty[16] or stent implantation[7]. Since the early '90s, this latter option has been proposed as a reliable alternative to surgery to achieve a stable duct patency[7-9] or to treat a shunt malfunction in high-risk patients[10-14]. In fact, this palliative approach is less invasive than surgery and enables precise fitting of the prosthesis to the size and angulations of the pulmonary arteries, thus avoiding any lung perfusion imbalance and pulmonary artery distortion. These peculiarities make do that the stented PDA is more effective and physiologic than a peripheral shunt of the same size. In addition, the transcatheter option, as compared to surgery, has the invaluable advantage of potentially allowing the titration of the shunt size to the pulmonary artery growth by repeat stent dilation[18,19,21]. However, early attempts at PDA stenting yielded discouraging outcomes due to the primitive techniques employed, the use of rigid and bulky sheaths, wires and guiding catheters, as well as the fact that the stent was not pre-mounted. Thus, owing to life-threatening complications, such as worsening cyanosis, bleeding, vessel rupture, duct spasm or acute stent thrombosis this approach was progressively abandoned[7,17]. Over time, technological improvements in coronary stent design and delivery systems have renewed interest in percutaneous techniques which have gained an important role as a reliable alternative to surgery, either as initial palliation of duct-dependent cardiac malformations or as a rescue option of surgical shunt malfunction.

In our series, the stenting procedure was carried out without any technical problems, mortality or significant morbidity. As previously reported[10-14,18-20], the early success rate and long-term efficacy of this procedure are critically dependent on the choice of the stent. In PDA stenting procedures, this should be long enough to cover the entire duct length, and particularly the pulmonary end that is its most reactive segment. The stent diameter should be individually tailored to the patient's size, as well as the pathophysiologic setting and the expected time of palliation. However, in our opinion, it should always be smaller than the indicated surgical shunt, in order to avoid any pulmonary overflow and/or competition with other blood flow sources. We believe that PDA stenting should be intended just as a short-term palliation until physiologic improvement is achieved or early corrective surgery performed. In this setting, it could be the ideal therapeutic option for patients with a pulmonary valve atresia and intact ventricular septum submitted to right ventricular radio-frequency decompression, in which often only short-term palliation is necessary to achieve biventricular physiology. Again, it could be advisable also in case of complex cardiac malformations with univentricular physiology destined to the Fontan operation, in which the stented ductus could just act as a bridge toward an early cavo-pulmonary anastomosis procedure. However, two important issues have so far not yet been completely addressed. First, the precise role of PDA stenting when a longer-term palliation must be provided is still unclear. Previous studies showed that the stented PDA was less durable than a conventional surgical shunt[17-20], due to duct tissue prolapse through the stent struts and elastic recoil of the stent consequent to contraction of the duct wall, although it allowed for an adequate pulmonary blood flow for several months, with a low risk of acute occlusion. Should a longer palliation be provided, the stent may be re-dilated over time to progressively increase the pulmonary blood flow, although the repeat passage of a previously stented vessel is quite troublesome and may result in more severe hemodynamic instability than the primary catheterization. Second, the data so far reported in the literature regarding the potential of the stented PDA in supporting the pulmonary artery growth as compared to the surgical shunt are scanty. This issue deserves further studies on larger series of patients before this option could be considered as a reliable alternative to the surgical shunt.

We believe that in contrast to PDA stenting, percutaneous treatment of a surgical shunt malfunction by stent implantation is no doubt preferable to the surgical revision or a second shunting procedure. Ideally, stent implantation should also be safer and more effective than balloon angioplasty, in that it avoids the elastic recoil of a kinked shunt conduit, as well as the distal dis-

lodgement of mural clots in case of partial shunt thrombosis. In this case, the stent length and diameter should be such as just to cover the stenotic segment and restore the prosthetic conduit to its original size, thus minimizing the blood-stent contact surface and reducing intrastent restenosis.

In conclusion, stent implantation is a technically feasible, safe and effective palliative option in high-risk surgical patients with congenital heart disease and reduced pulmonary blood flow. Following the introduction of improved materials and techniques, it has become less invasive than surgery and may be tailored to the clinical needs of the individual patient. Besides, owing to its durability, this treatment might be favored whenever a short-term palliation is necessary before a spontaneous improvement or early corrective surgery. Further studies should address its clinical impact on longer-term palliation, as well as in supporting pulmonary artery growth. However, the optimistic data reported in the current literature[18-20] allow for reasonable optimism regarding further extension of this therapeutic option even to elective and low-risk patients in the near future.

References

1. Alkhulaifi AM, Lacour-Gayet F, Serraf A, Belli E, Planche C. Systemic-pulmonary shunts in neonates: early clinical outcome and choice of surgical approach. Ann Thorac Surg 2000; 69: 1499-504.
2. Gladman G, McCrindle BW, Williams WG, Freedom RM, Benson LN. The modified Blalock-Taussig shunt: clinical impact and morbidity in Fallot's tetralogy in the current era. J Thorac Cardiovasc Surg 1997; 114: 25-30.
3. Norwood WI, Lang P, Hansen DD. Physiologic repair of aortic atresia-hypoplastic left heart syndrome. N Engl J Med 1983; 308: 23-6.
4. Tamisier D, Vouhe PR, Vermant F, Leca F, Massot C, Nerveux J. Modified Blalock-Taussig shunts: results in infants less than 3 months of age. Ann Thorac Surg 1990; 49: 797-801.
5. Fermanis GC, Ekangaki AK, Salmon AP, et al. Twelve-year experience with the modified Blalock-Taussig shunt in neonates. Eur J Cardiothorac Surg 1992; 6: 58-9.
6. Sachweh J, Dabritz S, Didilis V, Vazquez-Jimenez JF, Bernuth G, Messmer BJ. Pulmonary artery stenosis after systemic-to-pulmonary shunt operations. Eur J Cardiothorac Surg 1998; 14: 229-34.
7. Gibbs JL, Rothman MT, Rees MR, Parsons JM, Blackburn ME, Ruiz CE. Stenting of the arterial duct: a new approach to palliation for pulmonary atresia. Br Heart J 1992; 67: 240-5.
8. Kampmann C, Wippermann CF, Schmid FX. Transcatheter recanalization and stenting of the closed ductus arteriosus in duct dependent lung perfusion. Heart 1998; 80: 206-7.
9. Schneider M, Zartner P, Sidiropoulos A, Konertz W, Hausdorf G. Stent implantation of the arterial duct in newborns with duct-dependent circulation. Eur Heart J 1998; 19: 1401-9.
10. Zahn EM, Chang AC, Aldousany A, Burke RP. Emergent stent placement for acute Blalock-Taussig shunt obstruction after stage 1 Norwood surgery. Cathet Cardiovasc Diagn 1997; 42: 191-4.
11. Peuster M, Fink C, Bertram H, Paul T, Hausdorf G. Transcatheter recanalization and subsequent stent implantation for the treatment of early postoperative thrombosis of modified Blalock-Taussig shunts in two children. Cathet Cardiovasc Diagn 1998; 45: 405-8.
12. Peuster M, Fink C, Windhagen-Mahnert B, Paul T, Hausdorf G. Early postoperative dysfunction of a modified Blalock-Taussig shunt: successful therapy with angioplasty and stent implantation in the subclavian artery. Z Kardiol 2000; 89: 114-7.
13. El-Said HG, Clapp S, Fagan TE, Conwell J, Nihill MR. Stenting of stenosed aortopulmonary collaterals and shunts for palliation of pulmonary atresia/ventricular septal defect. Catheter Cardiovasc Interv 2000; 49: 430-6.
14. Vincent RN, Porter AG, Tam VK, Kanter KR. Diagnosis and catheter treatment of innominate artery stenosis following stage I Norwood procedure. Catheter Cardiovasc Interv 2000; 49: 415-8.
15. Rudolph AM, Heymann MA, Fishman N, Lakier JB. Formalin infiltration of the ductus arteriosus: a method for palliation of infants with selected congenital cardiac lesions. N Engl J Med 1975; 292: 1263-8.
16. Walsh KP, Abrams SE, Arnold SE. Arterial duct angioplasty as an adjunct to dilatation of the valve for critical pulmonary stenosis. Br Heart J 1993; 69: 260-2.
17. Gibbs JL, Uzun O, Blackburn ME, Wren C, Hamilton L, Watterson KG. Fate of the stented arterial duct. Circulation 1999; 99: 2621-5.
18. Gewillig M, Boshoff DE, Dens J, Mertens L, Benson LN. Stenting the neonatal arterial duct in duct-dependent pulmonary circulation: new techniques, better results. J Am Coll Cardiol 2004; 43: 107-12.
19. Alwi M, Choo KK, Latiff HA, Kandavello G, Samion H, Mulyadi MD. Initial results and medium-term follow-up of stent implantation of patent ductus arteriosus in duct-dependent pulmonary circulation. J Am Coll Cardiol 2004; 44: 438-45.
20. Michel-Behnke I, Akintuerk H, Thul J, Bauer J, Hagel KJ, Schranz D. Stent implantation in the ductus arteriosus for pulmonary blood supply in congenital heart disease. Catheter Cardiovasc Interv 2004; 61: 242-52.
21. Duke C, Rosenthal E, Qureshi SA. The efficacy and safety of stent redilatation in congenital heart disease. Heart 2003; 89: 905-12.