

CLINICAL STUDY

Stenting of stenotic aortopulmonary collaterals in non-surgically treated adolescent or adult patients with pulmonary atresia, ventricular septal defect and multiple aortopulmonary collaterals

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ABSTRACT

OBJECTIVES: To present our experience with stent implantations in non-surgically treated patients with pulmonary atresia, ventricular septal defect and stenotic aortopulmonary collaterals.

METHODS: Between April 2007 and June 2009, 6 stents in 4 patients were implanted into stenosed segments of MAPCAs. The median age and weight of patients at stent implantation were 24 years (range 13–34 years) and 58.5 kg (range 56–70 kg), respectively.

RESULTS: All implantations were successfully performed; there were no procedure associated complications. The median diameter increase of stenosed MAPCAs was 107 %. The mean arterial blood saturation increased from mean 78 % to 84 %. An early increase in the median exercise duration measured by 6MWT was 36 %. Neointimal in-stent fibroproliferation in 4/6 stents and stent fracture in 1/6 patients occurred in mid-term follow-up.

CONCLUSION: Percutaneous catheterization stent implantation into stenosed segments of MAPCAs is a palliative procedure, that might improve quality of life of non-surgically treated patients with PA, VSD, MAPCAs. Neointimal in-stent fibroproliferation and stent fracture can be expected as complications in mid-term follow-up. CT-angiography is recommended to confirm these complications in progressive satO₂ decrease (Fig. 3, Ref. 12).

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KEY WORDS: stent, PA, MAPCAs, non-surgically treated patients.

Introduction

Pulmonary atresia (PA) with ventricular septal defect (VSD) and multiple aortopulmonary collaterals (MAPCAs) is a complex cyanotic congenital heart defect with severe prognosis. Corrective surgery is indicated in all neonates with PA, VSD, MAPCAs. There is still a group of patients surviving without any surgical intervention, due to hypoplastic or absent native pulmonary arteries disabling surgical correction. Pulmonary blood perfusion is ensured by MAPCAs in these patients. MAPCAs are stenotic almost in 60% of cases. Stenoses are mainly located at the origin of MAPCAs, or at the site of confluence with native pulmonary arteries (if present). The stenoses can even progress with time. They result in decrease of lung perfusion, progression of cyanosis, and NYHA grade worsening. First ever tried treatment of stenosed MAPCAs was their balloon dilatation with poor outcome due to elastic recoil of the vessel (1). First ever reported successful balloon expandable stent implantation into stenosed MAPCAs was reported by

McLeod et al in 1994 (1). They reached exercise tolerance and arterial saturation improvement in short-term follow-up. The goal of our study was to evaluate our experience with stent implantations into the stenosed segments of MAPCAs in non-surgically treated adolescent or adult patients with PA, VSD, MAPCAs at the Children's Cardiac Center in Bratislava, Slovakia.

Methods

This case reports study represents a retrospective analysis of percutaneous stent implantations (n = 6) into stenosed MAPCAs in non-surgically treated (n = 4) adolescent or adult patients (3 females and 1 male) with PA, VSD and MAPCAs at our institution between April 2007 and June 2009. Balloon-expandable, premounted, stainless steel Palmaz Genesis (PG) stents (Cordis, Johnson & Johnson, Miami, FL, USA), premounted on 6–10 mm diameter OptaPro balloons (Cordis Endovascular, Warren, NJ, USA) were used in all procedures. For stenting, patients with focal stenoses of aortopulmonary collaterals were chosen. These stenosed segments were recognized either by CT – angiography or by diagnostic catheterization prior to interventional procedures. Proximal and distal invasive pressures of the stenosed segment were measured before stenting. The stenting was not performed in case of systemic or near systemic pressures distally. The diameters of balloons were chosen according to the maximum diameter of the

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non-stenotic part of MAPCAs. All patients were medicated with 3 months lasting clopidogrel (1 mg/kg p.o.) therapy and permanent acetylsalicylic acid (5mg/kg p.o.) therapy after intervention. The effect of stent implantation was evaluated in short-term follow-up as followed: 1. arterial satO₂ increase, 2. increase of vessel diameter, 3. increase of blood pressure distally to stenoses, 4. increase in physical exercise tolerance (6 MWT). Procedure-related complications were evaluated.

Possible complications as stent fracture (2) or in-stent neointimal proliferation (3) associated with arterial satO₂ decrease were evaluated in mid-term follow-up, since April 2007 to November 2013. Chest-X ray, CT – angiography (4, 5) or diagnostic catheterization were used for detection of these complications. Necessity and effects of re-interventions were also analysed.

Results

Case 1

19-years old male patient with non-surgically treated PA, VSD, MAPCAs clinically presenting with central cyanosis, arterial satO₂ 72 %, NYHA III, and recurrent syncope. CT-angiography delineated two segmentally stenosed MAPCAs (Fig. 1). PG2460PPS stent was implanted into right-sided MAPCA consequently (Fig. 2A). Arterial satO₂ increased from 64 % to 79 %. Post-stenting angiography confirmed markedly improved pulmonary blood flow to right lung (Fig. 2B). The minimal stenotic diameter of MAPCA



Fig. 1. Computed tomography angiography (VRT – reconstruction; posterior view) of bilateral MAPCAs with significant segmental stenoses.

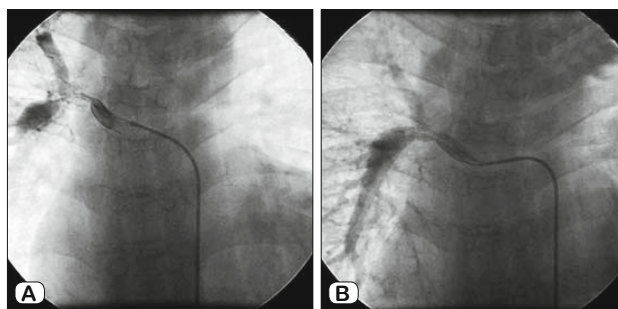


Fig. 2 Posteroanterior projection: (A) angiography of stenotic segment of right-sided MAPCA. (B) Posteroanterior projection: the final result after PG stent implantation with increased pulmonary blood perfusion in right lung segments.



Fig. 3. Computed tomography angiography (VRT – reconstruction; posterior view) after stent implantations into stenosed segments of MAPCAs.

increased from 2.5 mm to 5.5 mm. Pressures in MAPCA distally to implanted stent increased from 14/15/18 mm Hg to 28/27/26 mm Hg. 6 MWT walking distance increased from 180 m to 270 m. There were no intervention related complications. Implantation of next PG 2460PPS stent into left-sided MAPCA was performed following year. Arterial satO₂ increased from 76 % to 82 %. Post-stenting angiography confirmed markedly improved pulmonary blood flow to left lung. Effective lumen of MAPCA increased from 2.2 mm to 5.0 mm. Pressures in MAPCA distally to implanted stent increased from 14/13/13 mm Hg to 19/15/17 mm Hg. 6 MWT walking distance increased from 270 m to 400 m. There was no intervention related complication. Effective result of both stent implantations is shown in Figure 3. Progressive arterial satO₂ decrease to minimum 66 % was followed in mid-term follow-up. CTA revealed in-stent stenoses in both stents caused by neointimal fibroproliferation 5 years after first stent implantation. Angiography confirmed 4.5 mm effective lumen diameter in

right-sided stent and 4 mm effective lumen diameter in left-sided stent, consequently. Both stents were dilated by 8mm Opta Pro balloons (8–10 ATM). Stents shortened and widened to 6 mm diameter at the right side and 7 mm diameter at the left side. Subsequent angiography confirmed significant flow increase in both MAPCAs distally to implanted stents. Arterial satO₂ increased from 66 % to 76 %. Pressures in MAPCA distally to implanted stents increased from 14/8/11 mm Hg to 26/21/22 mm Hg at the right side, and from 18/10/14 mm Hg to 26/20/23 mm Hg at the left side. 6MWT distance increased from 200 m to 400 m. There were no intervention related complications. There were no syncope since stent implantations.

Case 2

33-years old female patient with non-surgically treated PA, VSD, MAPCAs clinically presenting with central cyanosis, arterial satO₂ 86 %, and NYHA III. CT-angiography delineated two segmentally stenosed MAPCAs and two non-stenosed MAPCAs. Two PG 2460PPS stents were implanted into both stenosed MAPCAs in 1 year lapse. Arterial satO₂ increased nonsignificantly from 86 % to 87 % after both procedures. Effective lumen of MAPCAs increased from 3.5 mm to 6.0 mm. There was no pressure change in MAPCA distally to implanted stent after intervention. 6 MWT could not be performed due to chronic neurological deficit (unilateral hemiparesis) after ischemic stroke. There was no intervention related complication. Progressive decrease of arterial satO₂ to minimum 75 % was followed during mid-term follow-up. CT-angiography revealed minimal in-stent stenoses of both stents caused by neointimal fibroproliferation 5 years after first stent implantation. Effective lumen diameters were 5.5mm in both stents. Both stents were dilated by 8 mm Opta Pro balloons (8–10 ATM). Subsequent angiography confirmed significant flow increase in both MAPCAs distally to implanted stents. Stents shortened and widened to 6.5 mm diameter at both sides. Arterial satO₂ increased from 75 % to 84 %. There was no intervention related complication.

Case 3

16-years old female patient with non-surgically treated PA, VSD, MAPCAs clinically presenting with central cyanosis, arterial satO₂ 82 %, and NYHA II–III. CT-angiography delineated one significantly stenosed MAPCA, and one nonstenotic MAPCA. PG 2510PPS stent was implanted into stenotic MAPCA consequently. Arterial satO₂ increased from 82 % to 92 %. Post-stenting angiography confirmed markedly improved pulmonary blood flow to right lung. Effective lumen of MAPCA increased from 4.8 mm to 8.0 mm. Pressures in MAPCA distally to the implanted stent increased from 14/10/12 mm Hg to 17/14/15 mm Hg. 6MWT walking distance increased from 450 m to 560 m. There was no intervention related complication. Arterial satO₂ 85–90 % persisted during 5 years follow-up.

Case 4

13-years old female patient with non-surgically treated PA, VSD, MAPCAs clinically presenting with central cyanosis, arte-

rial satO₂ 75 %, and NYHA II. CT-angiography delineated two significantly stenosed MAPCAs, both with very tortuous course. PG2510PPS stent was implanted into one MAPCA consequently. Second MAPCA was intended to be stented in the future. Arterial satO₂ 75 %, remained without any change. Effective lumen of MAPCA increased from 3.5 mm to 7.0 mm. Pressures in MAPCA distally to implanted stent increased from mean pressure of 13 mm Hg to 15 mm Hg. Chest X-ray and CT-angiography revealed complete fracture of implanted stent with preserved flow in distal part of MAPCA in 6 months follow-up. Effective diameter of stenotic MAPCA between fractured parts of PG stent was 3.5 mm again. Stabilization of fractured parts of PG stent by 30XL Andrastent (Andramed, Rutlingen, Germany) implantation mounted on OptaPro 12 mm/4 cm balloon (9–10 ATM) was performed consequently. As Opta 12 mm/4 cm balloon ruptured during inflation, stent dilatation with Opta 9 mm/2 cm balloon (12 ATM) was completed. Ruptured balloon was easily removed without any embolization. A central waist persisted on with comparison to 8 mm distal stent diameters. There was no change in arterial satO₂ 75 %, and no signs of stent fracture on chest X-ray in 5-years follow-up (6).

Results

Between April 2007 and June 2009, PG stents (n = 6) were implanted into stenosed MAPCAs in patients (n = 4) with non-surgically treated PA, VSD, MAPCAs. The patients ages ranged from 13 to 34 years (median 24 years), and their weight ranged from 56 to 70 kg (median 58.5 kg).

The mean arterial satO₂ increased from 78 % to 84 % (range 0–15 %). The mean MAPCA's diameter increased from 3 mm to 6.3 mm (range 2.5–4.0 mm). The mean blood pressure in distal MAPCAs increased from 13 mm Hg to 17 mmHg (range 2–11 mmHg). 6MWT median distance increased from 355 m to 455 m (range 20–130 m). There were no procedure associated complications.

There were stent associated complications with necessity of reinterventions in 5/6 (83 %) implanted stents during median 5 years (range 0.5–6 years) mid-term follow-up, since April 2007 to November 2013. Fractures of 1/6 (16.7 %) implanted stents and neointimal in-stent fibroproliferation of 4/6 (66.6 %) implanted stents presenting with cyanosis worsening were recognized during follow-up.

Successful implantation of 30XL Andrastent into fractured parts of previously implanted PG stent was performed 6 months after PG stent implantation with MAPCA widening from 3.5 to 5.0 mm in the narrowest part. There were no signs of stent fracture on chest X-ray in 5-years follow-up. The procedure was complicated by rupture of Opta 12 mm/4 cm balloon during inflation. Balloon was safely removed without any embolization.

Successful balloon dilations of 4 stenotic stents were performed in 2 patients. The median percentage of arterial satO₂ increase was 13.5 % (range 12.0–15.0 %). The median percentage of MAPCA enlargement was 24 % (range 14–55 %). There was 100 % walking distance increase in 6 MWT. We observed no procedure associated complications.

Discussion

Progressive stenoses of MAPCAs in non-surgically treated patients with PA, VSD, MAPCAs are leading to arterial satO_2 decrease. Stent implantation into stenosed segments of MAPCAs is a palliative procedure improving blood perfusion through lung segments supplied by these collaterals resulting in arterial satO_2 and exercise tolerance increase (7, 8, 9, 10, 11, 12). Redington et al reached significant arterial satO_2 increase from mean 64 % to 78 % and an early increase in exercise duration ($p < 0.01$) after self-expanding stents (Wallstent, Schnieder UK Inc) ($n = 12$) implantations in 11 patients (7). Brown et al presented arterial satO_2 increase from mean 75 % to 82 % (8) In comparison to these studies, we reached arterial satO_2 increase from mean 78 % to 84 %, and an early increase in exercise tolerance (6 MWT mean walking distance increase from 355 m to 455 m) after PG stents ($n = 6$) implantations in 4 patients. Minimal satO_2 increase in 2 of our patients can be explained by sufficient blood supply to the majority of lung segments through other untouched, non-significantly stenosed MAPCAs. We also reached mean MAPCA's diameter enlargement from 3 mm to 6.3 mm, and mean blood pressure increase from 13 mm Hg to 17 mm Hg in distal MAPCAs after stent implantation. We experienced no procedure associated complications. In comparison to our study Redington et al experienced two significant complications. Firstly, appearance of pink froth in endotracheal tube as a sign of pulmonary edema requiring positive-pressure ventilation for 36 hours. Secondly, compression of the branch of pulmonary artery by the stent with near occlusion. Patient necessitated surgical insertion of graft between right subclavian artery and right upper lobe artery (7).

We used stainless steel, balloon-expandable, closed-cell design PG stents in all procedures. Despite the technically demanding traverse through tortuous, stenotic MAPCAs, we could complete stents deployment in all procedures. In spite of that in Redington et al. study (7), and Brown et al study (8), they could not traverse through stenotic MAPCAs by a catheter in 1/13 and 2/25 stenoses, respectively. Thus occasional impossibility of stent implantation has to be considered in severely hypoplastic/stenotic MAPCAs. We do not recommend to perform stent implantations in non-stenotic or mildly-stenotic MAPCAs with mean distal pressures > 25 mm Hg due to high risk of pulmonary arterial hypertension accentuation in lung segments supplied by these vessels. Mean preprocedural distal MAPCAs pressure was 13 mm Hg in our study. Pulmonary arterial hypertension, as rare complication, can occasionally develop after stent implantation into significantly stenotic MAPCAs as it is reported by Redington et al (7). Mean postprocedural distal MAPCAs pressure was 17 mm Hg in our study. Extremely tortuous course of stenotic MAPCA could have been the reason for complete rupture of 1 stent in 6 months follow-up. Chest X-ray, CT-angiography and fluoroscopy could distinguish this complication (6). Successful implantation of 30XL Andrastent into fractured parts of previously implanted PG stent was performed consequently. Neointimal in-stent fibroproliferation presenting with cyanosis worsening can be expected as the

mid-term follow-up complication as we experienced in 4/6 stents in spite of prophylactic acetylsalicylic acid therapy. CT-angiography was an ideal imaging tool to visualize this pathology (4, 5). MRI is still inferior technique due to poorer spatial resolution and higher susceptibility to image artefacts. Redington et al observed minimal neointimal fibroproliferation (with thin rim < 1 mm) in 5/7 recatheterizations (7). Severe neointimal fibroproliferation or thrombus formation with significant cyanosis worsening was remarked only in 1/7 recatheterizations (7). The reason of this lower incidence of severe neointimal in-stent fibroproliferation in this study can be shorter follow-up after stent implantation (3 to 44 months) in comparison to median 5 years follow-up of our study. Neointimal in-stent fibroproliferations of MAPCAs in our study were successfully treated by catheterizational balloon dilatation with mean 13.5 % (range 12.0–15.0 %) satO_2 increase. Balloon redilatations can be expected due to recurrent neointimal in-stent fibroproliferation in the future.

Conclusion

Percutaneous catheterizational PG stent implantation into stenosed segments of MAPCAs is a palliative procedure, that might improve quality of life of non-surgically treated patients with PA, VSD, MAPCAs. We did not observe any significant procedure associated complications. Neointimal in-stent fibroproliferation and stent fracture can be expected as the complications in mid-term follow-up. CT-angiography is recommended to confirm these complications in progressive satO_2 decrease.

Learning points

- Perform stent implantations only in severely stenosed MAPCA
- Do not perform stent implantations in patients with sufficient blood supply to majority of lung segments through other untouched, non-significantly stenosed MAPCAs
- Do not perform stent implantations in non-stenotic or mildly-stenotic MAPCAs with mean distal pressures > 25 mm Hg
- You may expect neointimal in-stent fibroproliferation, or stent fracture in mid-term post-interventional follow-up
- Use CT-angiography to confirm these complications in progressive satO_2 decrease

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