Percutaneous treatment of stenosed major aortopulmonary collaterals with balloon dilatation and stenting: what can be achieved?

S C Brown, B Eyskens, L Mertens, M Dumoulin, M Gewillig

Abstract

Background—The natural history of major aortopulmonary collateral arteries (MAPCAs) in patients with pulmonary atresia and ventricular septal defect (PA-VSD) is frequently complicated by progressive stenosis, leading to pulmonary hypoperfusion and debilitating hypoxaemia.

Objective—To evaluate balloon dilatation and stenting for relief of stenoses and improvement of pulmonary flow in patients with PA-VSD.

Design—Retrospective analysis of all patients where dilatation of MAPCA stenoses was attempted.

Patients—Twelve patients with stenotic MAPCAs.

Interventions—Dilatation was attempted in 25 stenoses. Vessels were stented if elastic recoil was noticed (n = 3), in the presence of long segment stenosis (n = 4) or marked tortuosity (n = 1).

Main outcome measures—Diameter of stenoses before and after dilatation as well as arterial oxygen saturation data. Patients proceeding to surgical therapy.

Results—Two stenosed MAPCAs could not be crossed by a catheter. Four lesions were non-dilatable despite the use of high inflation pressures (18 atm). Six stenoses could be completely dilatated using angioplasty only; in five, only partial dilatation was obtained; eight stenoses needed stenting. In the group with partial expansion the mean (SD) diameter increased from 1.7 (0.8) to 3.5 (1.7) mm (p < 0.05); where full dilatation was achieved it increased from 2.1 (0.8) to 4.8 (1.9) mm (p < 0.05); and in the stented group it increased from 2.3 (0.9) to 5.0 (2.5) mm (p < 0.01). Percutaneous arterial oxygen saturation increased from 75(8)% to 82(8)% (p < 0.001). No complications were experienced during the procedures. Repeat dilatation was attempted in six stenoses, but only two procedures were successful. There were two episodes of vasospasm and in one an aneurysm had developed after redilatation. Two patients proceeded to outflow plasty and two subsequently had a unifocalisation procedure.

Conclusions—Pulmonary blood flow can be improved using balloon angioplasty or stents in patients with stenotic MAPCA; however, 17% of the lesions were not dilatable. Procedures are generally safe, but carry a small risk of vasospasm, dissection, occlusion or aneurysm formation.

(*Heart* 1998;**79**:24–28)

Major aortopulmonary collateral arteries (MAPCAs) in patients with pulmonary atresia and ventricular septal defects (PA-VSD) often have progressive stenosis resulting in pulmonary hypoperfusion.[1–4] The natural history of MAPCAs is, however, quite variable: in the same patient there may be multiple lesions, some stenotic that may progress to complete occlusion, and others without stenoses that usually become hypertensive.[3 4] There is a clear trend towards early unifocalisation or repair emerging in the literature.[6–9] However, patients with severely hypoplastic pulmonary arteries who do not qualify for correction are a difficult subgroup.[10]

The aim of this study was to assess the feasibility and safety of dilatating stenoses in MAPCAs and to assess the clinical effects on cyanosis, exercise ability, and facilitation of further surgical procedures.

Methods

PATIENTS

We reviewed all patients with MAPCAs who had undergone an interventional procedure to relieve stenosis. Balloon dilatation was limited to lesions accessible by catheter and was done to relieve critical stenoses or where there was fear of loss of lung segments. Patients with unifocalised MAPCAs or previous surgical repair were excluded. Twelve patients, six male and six female, had stenotic MAPCAs dilatated from July 1993 to November 1996. Age at the time of first procedure ranged from 1.5 to 32.2 years (mean age 12.5). The mean (SD) haemoglobin at the time of the procedure was 18.6 (2.2) g/dl (range, 15.5–22) and mean weight was 29.6 kg (range, 6.2–65).

There were 38 MAPCAs in the 12 patients with a mean of 3.2 collateral arteries per patient (range, 2–5). Five MAPCAs (13%) were hypertensive. Two vessels originated from the subclavian arteries, the remainder from the thoracic aorta.

PROCEDURE

Procedures were performed under general anaesthesia. Routine catheterisation data and measurements were obtained and pressure

Paediatric Cardiology, University Hospital Gasthuisberg, Leuven, Belgium
B Eyskens
L Mertens
M Dumoulin
M Gewillig

Paediatric Cardiology, University of the Free State, Bloemfontein, South Africa
S C Brown

Correspondence to:
Dr Gewillig, Paediatric Cardiology, UZ Gasthuisberg, Herestraat 49, B 3000, Leuven, Belgium. email: marc.gewillig@ uz.kuleuven.ac.be

Accepted for publication 29 September 1997

Figure 1 Example of MAPCA stenosis resistant to 20 atm. Notice the remaining waist of balloon (no dilatation at site of stenosis).

gradually withdrawn. On distal expansion of the stent, it was then "pulled" into place; the protective sheath was completely withdrawn resulting in full expansion of the stent across the stenosis.

Success of the procedure was assessed by angiography. If during balloon dilatation a residual waist remained in the balloon despite maximal safe inflation pressure, dilatation was considered to be incomplete. As we gained more experience, higher pressures were used. A stenosis was defined to be resistant if full expansion of the balloon was not obtained despite the use of high inflation pressures (18 atm) (fig 1).

Patients undergoing either angioplasty or stent placement were all given heparin during the procedure (50–100 U/kg, maximum 2500 U/h). No form of routine anticoagulation was initially given after stent placement; however, full anticoagulation was prescribed where low flow or significant residual narrowing was present after occlusion of a stent following redilatation.

FOLLOW UP
Chest radiography was done on all patients after 24 hours, followed after six weeks by clinical examination and repeat chest radiography. Thereafter the patients were seen according to clinical judgment.

ANALYSIS
The internal diameter of the narrowest segment before and after dilatation and stent implantation was measured. The values obtained before and after dilatation or stent implantation were analysed using the paired Student's t test; $p < 0.05$ was considered significant.

Results
Dilatation was attempted during 17 procedures in 25 MAPCAs. These were located in the proximal (n = 6), intermediary (n = 12), and hilar (n = 7) area.

Two stenoses could not be crossed either by a guidewire or the angioplasty balloon. In four MAPCAs no dilatation at the site of stenosis was achieved despite full inflation of the balloon and high inflation pressures of up to 18 atm (fig 1). Stenoses in these vessels were mostly located in the intermediary area of the MAPCA (n = 3). In all other stenoses, satisfactory balloon expansion was obtained with pressures less than 12 atm. Six of the stenoses were successfully dilatated by balloon angioplasty alone. Partial expansion of the balloon (with residual waist) was observed in five stenoses; no stent was then placed either because of a satisfactory clinical result, or because of the young age of the patient, where a stent would impede further growth of the MAPCA. Eight MAPCAs were stented for the aforementioned reasons. Eight stents were deployed in four patients in seven MAPCAs.

In the dilatation only group, mean diameter increased from 2.1 (0.8) to 4.8 (1.9) mm (p < 0.01), and from 2.3 (0.9) to 5 (2.5) mm (p < 0.005) in the stented collaterals (fig 2).

gradients repeated after stent implantation/balloon dilatation where possible. Standard techniques of balloon dilatation and stent placement were followed.[11 12] In short, stenoses were first crossed with a guidewire. If a critical stenosis was encountered, a 0.014" coronary guidewire was used and the vessel first dilatated by a small balloon (usually a 3 mm coronary balloon). An end-hole catheter was then advanced over the wire and a thicker (usually 0.035") guidewire left in situ for progressive dilatations or stent placement. All balloons were inflated by hand or with a pressure gauged syringe until disappearance of the waist. In some cases we exceeded the manufacturer's recommended burst pressure by up to 50%. Testing beforehand showed that the balloon (Ultra-thin ST, Medi-tech; Boston Scientific Corporation, Watertown, Massachusetts, USA) could safely withstand this pressure range without bursting. Choice of balloon size was made according to severity of stenosis and vessel size, but generally we selected a balloon not larger than the nearest normal vessel size.

Stents were used if immediate significant elastic recoil was noted after balloon angioplasty (n = 3), in the case of longer segment stenosis (n = 4) or in the presence of marked tortuosity of the collateral (n = 1). Choice of stent also varied according to shape, size, and length of stenosis, and whether there were acute angles proximal to the stenosis. All lesions were predilatated using appropriate balloons. Palmaz stents (Johnson & Johnson Interventional Systems, Warren, New Jersey, USA) were deployed using a long sheath placed beyond the stenosis. The stent mounted on a balloon was advanced through the sheath until it straddled the stenosis, and was expanded after withdrawal of the sheath. A Wallstent (Schneider, Bülach, Switzerland) was used if there was a small aorta with MAPCAs coming off at sharp angles, or in long stenoses or tortuous vessels. The mounted Wallstent was advanced over a 0.035" guidewire partially beyond the stenosis, and the protective sheath
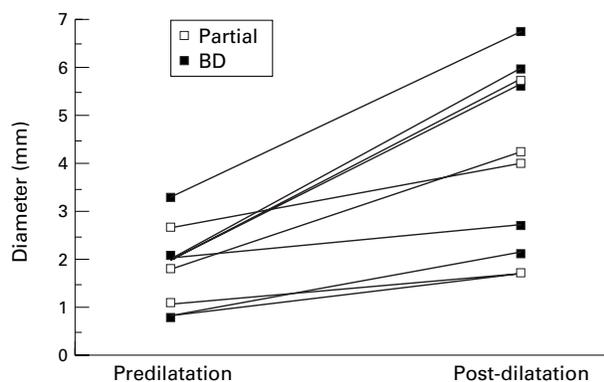
Figure 2   Change in diameter where partial balloon expansion was achieved (n = 5) and complete balloon dilatation (BD) (n = 6) before and after dilatation.
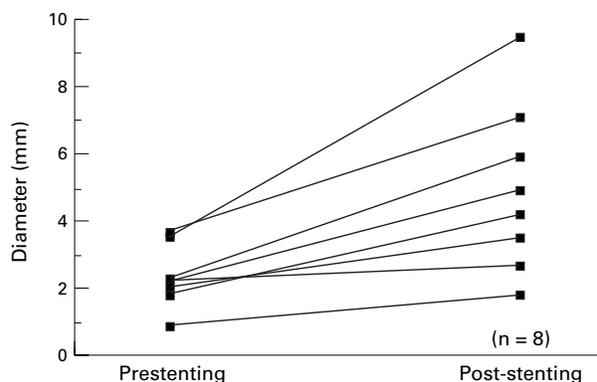

Figure 3   Increase in diameter of MAPCA stenoses before and after stenting.

Mean diameter increased from 1.7 (0.8) to 3.5 (1.7) mm (p < 0.05) in the group where partial expansion was achieved (fig 3).

Pressure gradients are available for only seven MAPCAs. Some stenoses were so severe that no predilatation measurements were possible; in others no postdilatation values were measured due to the risk of dislodging the stent or occlusion of the vessel. The systolic pressure in the distal pulmonary increased from 17 (4) (range, 12–25) to 40 (24) mm Hg (range, 14–65) (fig 4).

REDILATATION
Redilatation was attempted in six of the stenoses (two stents, four balloon dilatation only). In the four balloon dilatated stenoses, two were completely resistant to dilatation and one still had only partial expansion of the bal-

loon. One stent could not be redilatated even though a pressure of 20 atm was used. In one patient a proximal aneurysm developed after redilatation with pressures up to 18 atm. This patient subsequently developed a long dissection of the vessel wall with critical obstruction; two Wallstents were placed as an urgent rescue measure.

COMPLICATIONS
During dilatation and implantation no episodes of bleeding, perforation or rupture were experienced, and all stents were successfully deployed. There were two episodes of severe vasospasm during manipulation of the catheter in the vessels: one during primary balloon dilatation and one on redilatation. In the former the vessel was totally occluded on follow up catheterisation one year later. Residual angio-
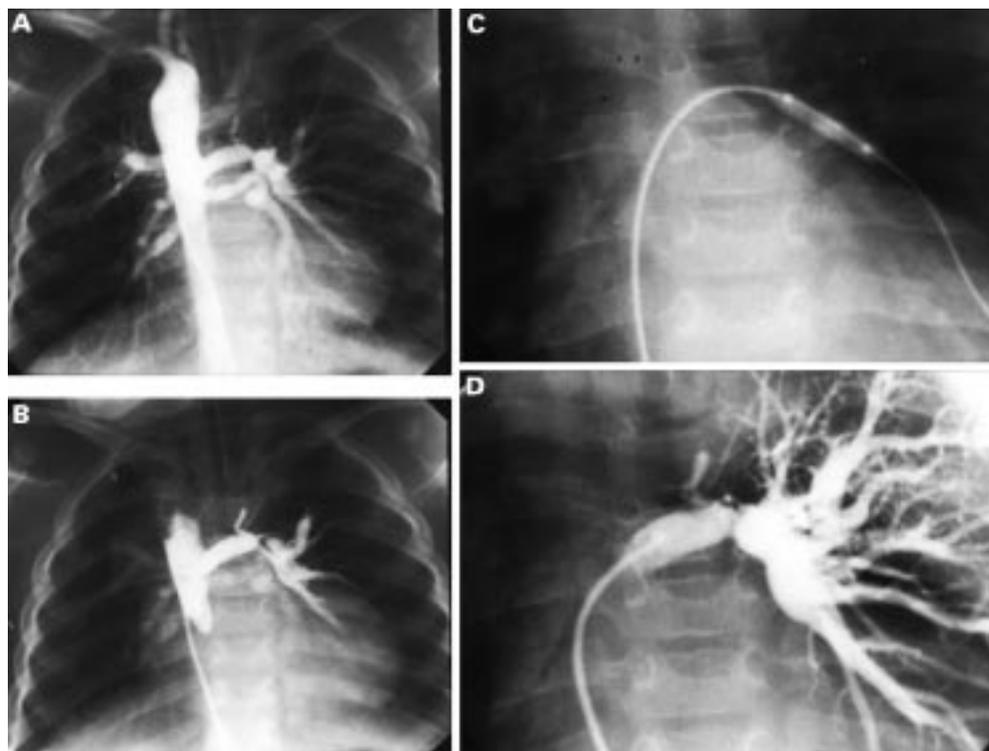

Figure 4   Angiogram (aortogram (A) and selective injection (B)) of a stenotic MAPCA perfusing the left upper lobe and lingula; note the poor peripheral perfusion. (C) Balloon dilatation: 6 mm balloon with complete disappearance of the waist. (D) Selective angiogram of the stenotic MAPCA after dilatation: there is better distal perfusion, aortic saturation had increased from 68% to 80% and distal mean pressure increased from 12 to 30 mm Hg.

graphic narrowing was present in three stents. In the stent group one patient had a pulmonary infarction of a lung segment due to occlusion of the MAPCA following redilatation. There was one sudden death 20 days after dilatation. The patient was severely cyanotic and symptomatic with both completely resistant and semicompliant stenoses.

FOLLOW UP

Patients have been followed for a mean (SD) of 2.1 (0.6) years (range, 0.6–2.8) after the first procedure. Before and after procedural percutaneous oxygen saturation data (room air) increased from a mean (SD) of 75(8)% (range, 65–85%) to 82(8)% (range, 68–87%). Three patients are in New York Heart Association class I, six in class II, and two in class III. Since the first procedure, two patients proceeded to a right ventricular outflow tract and two have had a modified Blalock-Taussig shunt connected to a previously dilatated MAPCA as part of a unifocalisation procedure.

Discussion

MAPCAs are embryologically preprogrammed not to survive beyond early fetal life. Should MAPCAs persist, there is a 58–68%[4] [13] incidence of significant stenoses in some series. Histologically, stenosis is produced by mounds of intimal tissue[4] [13] [14] that protrude into the lumen and contain significant amounts of collagen. The stenoses may progressively worsen due to ongoing turbulent flow, polycythaemia, hyperviscosity, and lack of growth.[1] [15] Patients will become progressively cyanotic, and lung vasculature may become progressively hypoplastic, with sometimes total occlusion and loss of the lung segment.

There is a clear trend towards early unifocalisation or repair emerging in the literature[6–9]; however, some lesions are not amenable to surgical procedures. Transcatheter interventions may therefore offer a valuable alternative.

In this series, some critical stenoses could not be probed with either a thin guidewire or a catheter. However, most stenoses could safely be accessed. Seventeen per cent of the stenoses were resistant to dilatation with pressures up to 18 atm. Such resistance is very uncommon in nature. Increasing the pressure beyond 12 atm never resulted in relief of stenosis, but was associated with more complications such as dissection or aneurysm of an adjacent vessel. However, the experience in this series is too small to be able to recommend not using pressures higher than 12 atm.

In the other MAPCAs, stenoses could significantly be relieved using balloon dilatation or stenting. This resulted in improvement of flow and satisfactory clinical palliation, with improved percutaneous oxygen saturations and clinical status. As with balloon dilatation of any vessel, there is a risk of vasospasm, dissection, aneurysm, occlusion, overdilatation, and rupture. The incidence, however, appears to be low provided balloons are kept small and short. Unfortunately no postmortem examination was performed on the patient who died, but there is no evidence linking death to the dilata-

tion procedure. This patient was severely symptomatic with extremely hypoplastic pulmonary arteries. Overdilatation should be avoided as four vessels became mildly hypertensive after the procedure. On follow up in two MAPCAs, pressures decreased in the distal pulmonary vasculature, predominantly because of growth of the distal vascular bed. One should be prepared to coil embolise a MAPCA should rupture occur. Hopefully, in future, covered stents could offer a safer bailout.

In this complex subgroup of patients, four proceeded to some form of surgery, which, before dilatation, was not believed to be possible. This indicates some beneficial effect of dilatation in selected patients. Growth of the peripheral vessels has been noticed after surgical restoration of flow[7] and we had the same impression on recatheterisation in patients where stenoses were successfully relieved.

Few problems have been encountered with the stents, but longer term follow up is needed to determine the outcome.[16] The indications for stenting as used in this study, namely elastic recoil, tortuosity, and longer segment stenosis, seem appropriate in the rehabilitation of collaterals. Initially we did not use routine anticoagulation after procedures, but more recently we have given anticoagulation for stents of smaller calibre and in low flow areas. This is in accordance with current literature[17] [18] where it seems appropriate to use anticoagulation in patients with smaller vessel diameter or partially expanded vessels.

LIMITATIONS

Several limitations of this study are recognised. The follow up is only short to medium term and long term data are required to determine the final outcome, especially as far as palliation is concerned. We also do not know, if surgery is preferred, to what extent dilatation of hilar stenoses will influence outcome and growth of the distal pulmonary vasculature. The lesions we dilatated were limited to a subgroup of patients not amenable to early surgical procedures. Furthermore, a wide variety and severity of stenoses in different locations of MAPCAs were treated, which could explain why some vessels occluded and others developed vasospasm or aneurysms.

CONCLUSIONS

Satisfactory clinical palliation can be achieved using percutaneous balloon dilatation and stenting of stenoses in MAPCAs with resultant improvement in pulmonary blood flow. In this series, 48% of lesions were dilatable using balloon angioplasty and 35% stretchable, but due to recoil and tortuosity placement of a stent was needed. Seventeen per cent of lesions were totally resistant to dilatation with pressures up to 18 atm. The procedure is generally safe, but carries a small risk of vasospasm, dissection, occlusion, and aneurysm formation, especially in the critical and more resistant stenoses. The increased pulmonary flow achieved after dilatation may create the opportunity for some surgical procedure in severely cyanosed patients.

1 Freedom RM, Rabinovitch M. Pulmonary atresia and ventricular septal defect. In: Freedom RM, Benson LN, Smallhorn JF, eds. *Neonatal heart disease.* London: Springer-Verlag, 1992:229–56.
2 Mair DD, Edwards WD, Julsrud PR, *et al*. Pulmonary atresia and ventricular septal defect. In: Emmanouilides GC, Allen HD, Riemenschneider TA, Gutgesell HP, eds. *Moss and Adams' heart disease in infants, children and adolescents,* 5th edn. Baltimore: Williams & Wilkins, 1995:983–97.
3 Rabinovitch M, Herrera-DeLeon V, Castaneda AR, *et al*. Growth and developement of the pulmonary vascular bed in patients with tetralogy of Fallot with or without pulmonary atresia. *Circulation* 1981;**64**:1234–49.
4 Haworth SG, Macartney FJ. Growth and developement of pulmonary circulation in pulmonary atresia with ventricular septal defect and major aortopulmonary arteries. *Br Heart J* 1980;**44**:14–24.
5 Ramsay MJ, Macartny FJ, Haworth SG. Tetralogy of Fallot with major aortopulmonary collateral arteries. *Br Heart J* 1985;**53**:167–72.
6 Shimazaki Y, Lio M, Nakano S, *et al*. Pulmonary artery morphology and hemodynamics in pulmonic valve atresia with ventricular septal defect before and after repair. *Am J Cardiol* 1991;**67**:744–8.
7 Reddy VM, Liddicoat JR, Hanley FL. Midline one-stage complete unifocalisation and repair of pulmonary atresia with ventricular septal defect and major aortopulmonary collaterals. *J Thorac Cardiovasc Surg* 1996;**109**:832–44.
8 Rome JJ, Mayer JE, Castaneda AR, *et al*. Tetralogy of Fallot with pulmonary atresia. Rehabilitation of diminutive pulmonary arteries. *Circulation* 1993;**66**:1691–8.
9 Shanley CJ, Lupinetti FM, Shah NL, *et al*. Primary unifocalisation for the absence of intrapericardial pulmonary arteries in the neonate. *J Thorac Cardiovasc Surg* 1993;**106**:237–47.
10 Hofbeck M, Sunnegardh JT, Burrows PE, *et al*. Analysis of survival in patients with pulmonic valve atresia and ventricular septal defect. *Am J Cardiol* 1991;**67**:737–43.
11 O'Laughlin MP, Perry SB, Lock JE, *et al*. Use of endovascular stents in congenital heart disease. *Circulation* 1991;**83**: 1923–39.
12 Perry SB, Keane JF, Lock JE. Interventional catheterisation in pediatric congenital and acquired heart disease. *Am J Cardiol* 1988;**61**:109G-17G.
13 Haworth S, Rees PG, Taylor JFN, *et al*. Pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries. Effect of systemic pulmonary anastomosis. *Br Heart J* 1981;**45**:133–41.
14 Thiene GT, Frescura C, Bini RM, *et al*. Histology of pulmonary arterial supply in pulmonary atresia with ventricular septal defect. *Circulation* 1979;**5**:1066–74.
15 Haworth SG. Collateral arteries in pulmonary atresia with ventricular septal defect. A precarious blood supply. *Br Heart J* 1980;**44**:5–13.
16 Redington A, Somerville J. Stenting of aortopulmonary collaterals in complex pulmonary atresia. *Circulation* 1996;**94**: 2479–84.
17 Moussa I, Mario CD, Francesco LD, *et al*. Subacute stent thrombosis and anticoagulation controversy: changes in drug therapy, operator technique, and the impact of intravascular ultrasound. *Am J Cardiol* 1996;**78**:13–17.
18 Kates MA, Jenkins S, Shaw D, *et al*. Endovascular stenting for treatment of an adult patient with pulmonary artery stenosis with one year follow-up. *Cath Cardiovasc Diag* 1996;**38**:417–20.