Percutaneous balloon valvuloplasty for pulmonary valve stenosis in infants and children

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SUMMARY Pulmonary valve stenosis was relieved by balloon dilatation during cardiac catheterisation on 27 occasions in 23 infants and children aged 7 days to 12 years, median 31 months (three aged <2 weeks). Pulmonary valve diameter was estimated by cross sectional echocardiography to assist in the choice of balloon size. Before dilatation the right ventricular systolic pressure ranged from 41 to 190 (median 92) mm Hg and was suprasystemic in 10 patients. There were significant reductions in the ratio of right ventricular to systemic systolic pressure and pulmonary systolic pressure gradients immediately after balloon dilatation. Twelve patients underwent recatheterisation (11 at six months and one at one week after balloon dilatation), which showed further improvement with significant reductions in right ventricular pressure or pulmonary valve gradient or both, particularly in the patients with the least satisfactory initial results. This improvement was attributed to resolution of the obstruction at infundibular level. Repeat pulmonary valve dilatation was successfully performed in four patients who had poor results after initial dilatation.

Balloon pulmonary valvotomy appears to provide good short term and medium term relief of pulmonary valve stenosis and may obviate the need for surgery in many cases. An apparently poor immediate haemodynamic result does not preclude a good longer term result.

The relief of pulmonary valve stenosis by balloon dilatation during cardiac catheterisation was first reported in 1982.¹ Subsequently, the results of this procedure have been reported in series of patients with wide age ranges (3 months to 56 years)²⁻⁴ and even in a neonate.⁵ Good immediate results were obtained in each of these studies.

Spontaneous regression of residual right ventricular outflow systolic pressure gradients and right ventricular systolic pressure has been reported after surgical pulmonary valvotomy.⁶ This is presumably related to regression of right ventricular infundibular hypertrophy. A similar phenomenon has not previously been reported after balloon dilatation of the pulmonary valve in children.

The aim of this study was to determine which

children from a consecutive series with a wide range in age and severity of pulmonary valve stenosis would respond favourably to balloon dilatation and whether this response would be maintained.

Patients and methods

The relief of pulmonary valve stenosis by balloon dilatation was attempted on 27 occasions in 23 patients between April 1984 and February 1985. Patients were aged 7 days to 12 years (median 31 months). Three patients were aged <2 weeks (Table).

Eight patients with isolated pulmonary valve stenosis were catheterised, but balloon dilatation not performed, during the same period. In three neonates with severe pulmonary valve stenosis satisfactory catheter placement across the pulmonary valve could not be obtained. In three others crossing the stenosed valve with a balloon catheter was not considered prudent. In two patients, investigated early in the series, the peak pressure in the right

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Table Systolic pressures (mm Hg) at rest, immediately after balloon pulmonary valve dilatation, and at repeat catheterisation

Case No	Age	Before dilatation			After dilatation			At repeat catheterisation		
		RV	PA	Sys	RV	PA	Sys	RV	PA	Sys
Typical pulmonary valve stenosis										
1	2 y	95	16	94	58	19	105	33	18	90
2	6 y	100	28	104	55	38	103	48	30	105
3	18 mth	70	15	114	55	20	100	53	33	130
4	6 y	125	20	120	57	20	100	40	14	107
5	2 y	75	18	95	46	25	80	47	21	101
6	5 y	164	11	93	86	24	80	33	21	96
7	14 mth	68	21	86	43	15	92			
8	22 mth	144	19	70	140	23	100	72	10	85
					48*	23*	95*			
9	2 y	66	24	87	29	21	98			
10	8 d	165	20	118	127	31	113	92	32	83
					58*	35*	81*			
11	19 mth	87	16	95	43	20	104			
12	12 d	70	40	80	53	43	81			
13	2 y	41	15	96	44	32	96			
14	12 y	51	14	126	43	21	112			
15	17 mth	67	32	90	67	32	100			
16	7 y	190	26	108	72	20	108			
17	7 d	88	54	64	37	34	49			
18	18 mth	83	14	74	40	14	73			
19	10 y	107	24	121	38	21	109			
Noonan's syndrome										
20	11 y	65	24	100	55	25	108	60	15	105
21	5 y	85	15	106	69	14	106	60	24	106
					35*	20*	113*			
Previous surgical valvotomy										
22	3 y	120	28	97	122	28	103	62	25	98
23	4 y	116	21	104	96	25	98	90	17	108
					43*	23*	110*			

*Pressures after repeat balloon dilatation. PA, pulmonary artery; RV, right ventricle; sys, systemic.

ventricle was twice that in the left ventricle. The third patient had suprasystemic pressure in the right ventricle and had arrested after ventriculography. Surgical valvotomy was performed in these six patients. The severity of pulmonary valve stenosis was insufficient to indicate balloon valvotomy or surgery in an infant weighing 3.5 kg and in an older child with Noonan's syndrome. Balloon valvotomy was not performed when pulmonary valve stenosis was associated with a large atrial septal defect in two patients as it would not obviate the need for surgery.

INTERNAL CARDIAC MEASUREMENTS

The diameter of the pulmonary valve ring was estimated by cross sectional echocardiography before catheterisation. Internal dimensions were measured by electronic calipers on video freeze frames, where the origins of the pulmonary valve leaflets could be clearly seen, without specific regard to the phase of the cardiac cycle. Multiple views were used, but short axis parasternal cuts were usually used to determine the choice of balloon size. External balloon diameters that were equal to or slightly less than the measured pulmonary valve ring diameter were selected for the first 14 procedures: balloon diam-

eters about 20% greater than the pulmonary valve ring diameter were used subsequently except in one patient (case 14) (Table), in whom the largest balloon diameter available (20 mm) was 5 mm less than the measured pulmonary valve ring diameter.

CARDIAC CATHETERISATION

Cardiac catheterisation was performed under local anaesthesia usually through a saphenous vein cut down or a femoral vein after percutaneous puncture. A right axillary cut down was used in one patient after multiple previous investigations. Oximetry and haemodynamic measurements were recorded at rest, which always included right ventricular and pulmonary artery pressure, and were followed by right ventricular angiography. Femoral artery cannulation allowed continuous monitoring of the systemic arterial pressure, although this was not performed in the neonatal patients. A balloon catheter (Medi-Tech) of appropriate size was then introduced and positioned across the pulmonary valve using the appropriate exchange guide wire previously introduced into the left pulmonary artery, or into the descending aorta via a persistent ductus arteriosus in two neonates. The lateral image intensifier screen was the preferred

Fig. 1 Lateral right ventricular angiogram in a child with pulmonary stenosis (a) before and (b) after dilatation and (c) at repeat study. (b) After partial inflation of the balloon indentation by the pulmonary valve leaflets (pv) is present, which disappears after full inflation (c). PA, pulmonary artery; RV, right ventricle; B, balloon; pv, pulmonary valve; A, anterior; S, superior.

plane for determining satisfactory balloon placement. Rapid balloon inflation with dilute contrast medium was performed by hand, ideally until the indentation in the partially inflated balloon caused by the pulmonary valve leaflets was overcome (Fig. 1). Inflation was maintained for about 10 s, which usually coincided with a rapid decrease in systemic arterial pressure. Systemic pressure returned to normal with deflation of the balloon. Balloon inflation was repeated three to five times in each case. The balloon catheter was then re-exchanged for

a Goodale-Lubin catheter for repeat haemodynamic measurements, and right ventricular angiography was also repeated with a suitable catheter.

Repeat cardiac catheterisation has been performed in 12 patients to date. In 11 patients this was an elective repeat study six months after the initial procedure. Repeat balloon dilatation was performed in three of these patients. One neonate with severe pulmonary valve stenosis (case 10) was recatheterised one week after the initial procedure and repeat balloon dilatation performed.

Fig. 2 (a) Right ventricular to systemic systolic pressure ratios and (b) pulmonary systolic pressure gradient before and immediately after balloon pulmonary valve dilatation. Bars represent mean (SD).

Results

The Table shows the patients' characteristics. Nineteen patients had isolated pulmonary valve stenosis, although one of these (case 15) had an associated right coronary artery to right ventricle fistula. There were two patients with Noonan's syndrome, and two who had undergone previous surgical valvotomy. The only complication of cardiac catheterisation in these patients was avulsion of the saphenofemoral junction in one neonate, which necessitated femoral vein ligation. All patients, except the three aged < 2 weeks, were discharged from hospital the day after catheterisation.

INITIAL DILATATION

Ten patients had suprasystemic right ventricular pressure before dilatation and 21 of the 23 had right ventricle to systemic systolic pressure ratios > 0.6 (Fig. 2). One patient (case 13) did not have a large pulmonary valve gradient, but balloon dilatation was

Fig. 3 (a) Right ventricular to systemic systolic pressure ratios and (b) pulmonary systolic pressure gradients before and immediately after balloon pulmonary valve dilatation and at repeat catheterisation.

Fig. 4 Right ventricular lateral angiograms (a) before (b) immediately after balloon pulmonary valve (PV) dilatation and (c) at repeat catheterisation, showing regression of infundibular (Inf) stenosis. PA, pulmonary artery; RV, right ventricle.

performed in an attempt to reduce the moderate tricuspid regurgitation that was present.

Balloon pulmonary valve dilatation resulted in significant immediate haemodynamic improvement. Mean (SD) right ventricular to systemic systolic pressure ratios and peak to peak pulmonary systolic pressure gradients were 1.02 (0.42) and 76 (41) mm

Hg respectively before dilatation compared with 0.67 (0.29) ($p < 0.001$) and 39 (31) mm Hg ($p < 0.001$) after balloon dilatation (Fig. 2). Right ventricular systolic pressure remained at or above systemic level on five occasions, however (Fig. 2a), and in four of these patients the residual pulmonary systolic pressure gradient was > 70 mm Hg (Fig. 2b). These

patients (cases 6, 8, 10, 22, and 23) who had had the least satisfactory initial results underwent a repeat study.

LATE RECATETERISATION

There was further appreciable haemodynamic improvement in the 12 patients who underwent repeat study (Fig. 3). Right ventricular to systemic systolic pressure ratios and pulmonary systolic pressure gradients for this group were 0.8 (0.32) and 56 (33) mm Hg respectively immediately after dilatation, compared with 0.58 (0.23) ($p < 0.01$) and 36 (20) mm Hg ($p < 0.05$) at repeat study. This further improvement was modest in patients who had the lowest right ventricular pressures and residual gradients after the initial balloon dilatation procedure. More notable, however, was the major improvement with time that had occurred in the patients who had had less satisfactory initial results (Fig. 3). Four patients with high residual pulmonary gradients and right ventricular systolic pressures underwent successful repeat dilatation, in each case with a bigger balloon than on the first occasion (Fig. 3). Repeat dilatation could not be achieved in one patient (case 22) whose right ventricular pressure remained at 63% of systemic pressure because the balloon catheter could not be advanced through a left axillary cut down. This patient had undergone surgical valvotomy with a modified Blalock-Taussig shunt on the first day of life for pulmonary atresia with intact ventricular septum. He had a severely dysplastic valve. Although there was no immediate improvement after balloon dilatation, a major improvement did occur six months later. The only other patient with a residual gradient in excess of 26 mm Hg (case 20) or right ventricular pressure more than half systemic pressure was a child with Noonan's syndrome and multiple handicaps. In view of the latter and the lack of benefit from the initial dilatation in a child with moderate stenosis only, we elected not to perform repeat balloon dilatation.

Discussion

Our results confirm earlier reports that balloon dilatation of the pulmonary valve may provide effective relief of pulmonary valve stenosis. Moreover, this is not confined to pulmonary valve stenosis of mild or moderate severity since almost half of the patients in this series had suprasystemic right ventricular pressures. Previous reports of balloon dilatation for pulmonary valve stenosis of similar severity included older patients (age range from 11 months to 19 years), with the most severe lesions occurring mainly

in the older patients,^{3,4} but we have applied the technique to infants with critical pulmonary stenosis.

The pronounced spontaneous haemodynamic improvement that we noted in those patients with the least satisfactory initial haemodynamic results has not been reported previously. This occurred in patients with the most severe initial gradients and right ventricular hypertension. The pronounced residual obstruction in these patients was probably at infundibular level, the subsequent improvement being associated with regression of right ventricular hypertrophy (Fig. 4). We were unable to show separate residual gradients at valvar and infundibular levels after the initial dilatation, however, despite withdrawal of an end hole catheter from the pulmonary trunk to the right ventricle. Direct trauma to the infundibulum from the inflated balloon may have exacerbated the gradient immediately after dilatation.⁷ A previous study has shown that reductions in systolic pressure gradients and right ventricle to systemic systolic pressure ratios were maintained, but not further improved, in nine patients with typical pulmonary valve stenosis studied again two to 12 months after the initial procedure.² Two single cases of resolution of mild residual infundibular gradients after balloon pulmonary valvotomy in adults (one of whom had only mild pulmonary stenosis) have been reported.^{8,9} Analogous resolution of infundibular obstruction has also been reported after surgical pulmonary valvotomy.⁶ The administration of 0.1 mg/kg of propranolol had no effect on right ventricular pressure immediately after balloon pulmonary valve dilatation in one patient (case 6) (Fig. 4) and was not therefore used routinely. Hence an apparently poor immediate result from balloon pulmonary valve dilatation does not preclude a later good result.

Early in our experience we used balloons that were probably too small for optimal results. Studies in lambs suggest that the optimum balloon diameter should be about 20–30% greater than the pulmonary valve ring diameter,⁷ but if the balloon diameter exceeds 150% of the pulmonary valve ring diameter severe damage to the right ventricular outflow tract, particularly the infundibulum, can occur. Support for this is provided by the effectiveness of repeat dilatation in the four patients in whom a larger balloon was used. None of the patients whom we studied again had a diastolic murmur to suggest pulmonary incompetence.

There was a higher proportion of infants in this series than in previous studies, although our technical failures occurred in neonates and small infants. This resulted partly from the relatively large size of the catheter required to carry the appropriate sized balloon in these small patients. This contributed to

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avulsion of the saphenofemoral junction in one neonate, the only complication of the procedure encountered. Balloon length and stiffness of the catheter tip also made manipulation difficult in neonates. Although catheter design is likely to improve, the technical problems of entering the pulmonary artery with these large stiff catheters may not allow all symptomatic neonates with severe pulmonary valve stenosis to be improved without surgery.

Poor results from balloon dilatation of pulmonary valve stenosis in patients with dysplasia of the pulmonary valve or previous neonatal surgical valvotomy have been reported.² We had a similarly unsatisfactory result in one patient with Noonan's syndrome, although, in retrospect, the balloon used was probably too small. There was a good result (after two dilatation procedures), however, in another patient with Noonan's syndrome. As patients with dysplastic pulmonary valves are unlikely to form a homogeneous group, this suggests that percutaneous balloon dilatation may still be the initial treatment of choice. We have no means yet of identifying patients with optimal pulmonary valve anatomy. Both children in this series with severe residual pulmonary stenosis after surgical valvotomy (performed at 1 day and 6 months of age respectively) had major haemodynamic improvement with balloon valvuloplasty, albeit after two dilatation procedures in one child. Previous surgical valvotomy is not, therefore, a contraindication for this technique. Stretching of fibrous tissue at the pulmonary valve ring may have contributed to this improvement. This may be analogous to successful balloon dilatation of postoperative residual coarctation of the aorta.¹⁰

Balloon pulmonary valvuloplasty did not abolish the systolic pressure gradient in any of our patients, although these results are at least similar to those after surgery, where about 20% of infants and children studied again postoperatively had a residual gradient >25 mm Hg.¹¹ The same study showed that pulmonary systolic pressure gradients of <30 mm Hg were unlikely to increase, whether they occurred in patients managed medically or in those with a residual gradient after surgery.¹¹ Residual gradients after balloon pulmonary valvuloplasty therefore, will probably behave similarly, although this remains to be proved.

Addendum

Since this paper was prepared further studies on this subject have been published.^{12,13}

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