TECHNIQUE

Initial experience with balloon dilatation of supralvalvar aortic stenosis

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Abstract

Primary balloon dilatation of supralvalvar aortic stenosis was attempted in three patients: a 20 year old woman, a seven month old boy, and a 12 year old girl. Balloon catheters (Mansfield) with diameters of 25 mm, 8 mm, and 15 mm were used in the three patients respectively. The systolic pressure gradient across the aortic narrowing decreased considerably and the diameter of the constricted area increased significantly.

Balloon dilatation was feasible and provided good immediate results and sustained relief of supralvalvar aortic stenosis in these three patients. However, a larger study is needed to establish the place of this procedure in treatment.

(Br Heart J 1993;70:476-478)

Supralvalvar aortic stenosis is the least common cause of obstruction of the left ventricle outflow tract. It is caused by narrowing of the ascending aorta just above the aortic sinuses. There are three anatomical types: (a) hour-glass, (b) hypoplastic and (c) membranous.

The good results reported with balloon dilatation of valvar,^{1,2} subvalvar,³⁻⁵ and supralvalvar aortic stenosis,⁶ prompted us to attempt balloon dilatation of supralvalvar aortic stenosis in three patients.

Patients and methods

Patient 1 was a 20 year old woman, patient 2 was a seven month old boy, and patient 3 was a 12 year old girl. None had classic clinical signs of William's syndrome. Patients 1 and 3 had dyspnoea on exertion and patient 2 had dyspnoea caused by crying or sucking.

All patients had clinical features of supralvalvar aortic stenosis, including systolic

murmur and thrill in the aortic area with radiation to the suprasternal notch and carotid arteries.

The electrocardiogram showed left ventricular hypertrophy in patients 1 and 3 and right ventricular hypertrophy in patient 2. There were no associated abnormalities in any of the three patients.

Supralvalvar aortic stenosis was diagnosed by cross sectional echocardiography. The systolic pressure gradients measured by Doppler echocardiography were 78 mm Hg, 72 mm Hg, and 104 mm Hg in patients 1, 2, and 3 respectively. The pressures in the pulmonary artery and right ventricle derived from the modified Bernoulli formula or by the flow in pulmonary artery (Graettinger formula) were normal in all patients.

Patients were sedated with diazepam or meperidine and cardiac catheterisation was performed through the percutaneous femoral artery approach in patients 1 and 3 and by femoral arteriotomy in patient 2.

Pressures in left ventricle and aorta were recorded through a NIH angiographic catheter. An aortogram and left ventriculogram obtained in left anterior oblique view showed supralvalvar narrowing. Though other angiographic views can be used to measure the aortic diameter we prefer this view because it shows the opacified ascending aorta, aortic isthmus, and the thoracic descending aorta at the same time. If the portion of the ascending aorta above the stenosis had a diameter similar to the aortic isthmus it was regarded as normal. We calculated the diameter of the normal ascending aorta using a magnification factor for correction.

A 0.038 inch flexible tip guide wire that was 260 cm long (Cook) was passed through a pigtail catheter and advanced into the left ventricle. The pigtail catheter was withdrawn and a balloon catheter (Mansfield) (diameter 25 mm in patient 1, 8 mm in patient 2, and 15 mm in patient 3) was inserted over the guide wire and positioned across the narrowing and inflated three or four times, without use of manometer for 10-15 seconds.

To reduce the risk of aneurysm formation or rupture of the aortic wall we never used a balloon catheter with a diameter bigger than the diameter of the normal aorta. In patients 2 and 3 we used balloons that were 1 mm smaller than the aortic diameter. Because in

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Accepted for publication 21 July 1993

Systolic gradient and diameter of the constriction before and immediately after balloon dilatation

Patient	Gradient (mm Hg)			Diameter (mm)		
	Before	After	Decrease (%)	Before	After	Increase (%)
1	64	27	57.8	13	18.5	42
2	65	15	76.9	4.5	8.0	77
3	102	40	60.7	9	13	45

Figure 1 Aortic angiograms obtained before and after dilatation in patients 1 (A and B), patient 2 (C and D), and patient 3 (E and F).

patient 1 the aorta above the stenosis was dilated we used a balloon with a diameter similar to the aortic annulus.

The guide wire was kept in position and the balloon catheter was replaced with the pigtail catheter. The guide wire was then removed and pressures in the left ventricle and aorta were recorded.

Repeat aortograms and left ventriculograms were obtained in left anterior oblique view.

Results

In all cases a systolic gradient was detected

above the aortic valve. Mild (grade 1) aortic valve regurgitation was demonstrated angiographically in all the three patients before balloon dilatation. It did not increase after the procedure. All the three patients had hour-glass aortic narrowing.

Left ventricular function was normal before and after the procedure in all patients. The table shows the gradient across the supravulvar stenosis and the diameter of the constricted segment before and after balloon dilatation.

Figures 1 and 2 show the angiographic images and the pressure recorded before and after balloon dilatation.

Figure 2 Aortic pressures recorded by withdrawal of the catheter through the aorta before and after dilatation in patient 1 (A and B), patient 2 (C and D), and patient 3 (E and F). Numbers above the pressure curves are systolic aortic pressures in mm Hg.

Patients were discharged 48 hours after the procedure. There were no procedure-related complications but the infant (patient 2) died 30 days after the procedure from an infectious disease. A necropsy was not performed. Dyspnoea was relieved in all patients. Patients 1 and 3 remain symptom free. In patient 1 the Doppler systolic pressure gradient was 34 mm Hg 22 months after dilatation and in patient 3, it was 50 mm Hg three months after dilatation. In our experience the Doppler gradient is always about 20% bigger than the gradient recorded during catheterisation.

Discussion

Balloon dilatation is a new and promising procedure for the treatment of aortic coarctation.⁷⁻⁹ It has been used to treat stenosis caused by aortitis.¹⁰ Our experiences with these three patients indicates that fibromuscular narrowing of the aorta is amenable to balloon dilatation.

Surgical treatment of supravalvular aortic stenosis by patch aortoplasty is well established and gives good postoperative results. Residual abnormalities may remain, however, including aortic valve insufficiency and residual outflow tract obstruction.¹¹

We found that balloon dilatation relieved supravalvular aortic stenosis. There was a considerable fall in the systolic gradient and the diameter of the constricted area of the aorta increased significantly.

Even in the patient who was 20, the result was good and the late follow up showed sustained relief of stenosis. Dilatation was reported to be impossible in another 20 year

old patient, probably because the lesion was rigid.⁶

The results in our three patients suggest that balloon dilatation of supravalvular aortic stenosis, gave good results with few little psychological or physical side effects and early discharge from hospital. We need to study more patients for longer follow up to determine the place of this procedure in treatment.

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