The dysplastic pulmonary valve: echocardiographic features and results of balloon dilatation

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SUMMARY The feasibility of using balloon dilatation to relieve stenosis caused by dysplasia of the pulmonary valve was assessed in seven patients (five female, mean age two years) with angiographically confirmed dysplasia who were identified among 38 patients with pulmonary valve stenosis selected for balloon dilatation over a two year period. The clinical features in three patients were consistent with Noonan's syndrome. In all patients the gradient across the valve was assessed by cross sectional echocardiography and Doppler echocardiography before cardiac catheterisation. Balloon dilatation was performed by conventional techniques. In one patient, who had balloon dilatation in the operating room before surgical valvectomy, the diameter of the valve orifice increased from 3 mm to 10 mm. Inspection showed a tear along the anterior commissure. The mean (SD) pressure gradients between the right ventricle and pulmonary artery before and immediately after dilatation in five patients were not significantly different (58 (28) and 47 (13) mm Hg) respectively. There was no overall significant change in the degree of stenosis when four of these patients were examined by Doppler echocardiography six months after operation (44 (12) mm Hg), although one patient (case 5) did show a significant reduction in gradient. This patient had angiographic and echocardiographic features of dysplasia and commissural fusion. Several echocardiographic features were common to all patients and distinguished them from cases of typical pulmonary valve stenosis. These were: (a) pronounced thickening of leaflets; (b) leaflet immobility in diastole and systole; (c) no dilatation of the sinuses of Valsalva in diastole, and (d) supra-annular narrowing.

These poor results of balloon dilatation suggest that commissural fusion is not an important mechanism for causing stenosis in the dysplastic pulmonary valve. When dysplasia of the pulmonary valve is identified clinically and echocardiographically, balloon dilatation is unlikely to improve haemodynamic function; however, it should be attempted if commissural fusion is present.

About 10% of cases of congenital heart disease have isolated pulmonary valve stenosis.1 The mechanism of stenosis may be commissural fusion, resulting in a reduced haemodynamic orifice, or dysplasia of valve leaflets,2-6 which presents obstruction to flow, with or without a commissural component and a hypo-

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Table 1  Results of balloon dilatation in seven patients with dysplastic pulmonary valves

<table>
<thead>
<tr>
<th>Patient No</th>
<th>Annulus size (mm)</th>
<th>Pre-dilatation</th>
<th>Post-dilatation</th>
<th>Follow up</th>
</tr>
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<tbody>
<tr>
<td></td>
<td></td>
<td>RV pressure as % of systemic</td>
<td>Gradient (mm Hg)</td>
<td>RV pressure as % of systemic</td>
</tr>
<tr>
<td>1</td>
<td>15</td>
<td>90</td>
<td>60</td>
<td>—</td>
</tr>
<tr>
<td>2</td>
<td>10</td>
<td>69</td>
<td>36</td>
<td>—</td>
</tr>
<tr>
<td>3</td>
<td>11</td>
<td>60</td>
<td>40</td>
<td>67</td>
</tr>
<tr>
<td>4</td>
<td>11</td>
<td>142</td>
<td>150</td>
<td>—</td>
</tr>
<tr>
<td>5</td>
<td>10-4</td>
<td>103</td>
<td>96</td>
<td>70</td>
</tr>
<tr>
<td>6</td>
<td>8</td>
<td>86</td>
<td>80</td>
<td>72</td>
</tr>
<tr>
<td>7</td>
<td>10</td>
<td>60</td>
<td>38</td>
<td>60</td>
</tr>
<tr>
<td>Mean (SD)</td>
<td>10-7 (2.11)</td>
<td>87 (29)</td>
<td>71.4 (41.5)</td>
<td>63.8 (8.9)</td>
</tr>
</tbody>
</table>

RV, right ventricular.
†Only patients dilated.

Percutaneous balloon dilatation has now replaced open heart valvotomy for the relief of typical pulmonary valve stenosis.12-17 The attendant morbidity is minimal and short term results appear to be good. A few patients with dysplasia of the pulmonary valve have undergone this procedure but balloon dilatation has not been specifically assessed in this condition.14 15 17 We report our experience of using balloon dilatation in patients who had dysplasia of the pulmonary valve that was identified by cross sectional echocardiographic criteria.

Patients and methods

Seven of 38 patients undergoing percutaneous balloon dilatation of the pulmonary valve between January 1984 and December 1985 were prospectively identified as having clinical features consistent with dysplasia of the pulmonary valve. Three patients had the Noonan phenotype (cases 1, 2, and 3). None had a pulmonary ejection click. Plain chest radiographs did not reveal post-stenotic dilatation of the main pulmonary artery (a feature noted in at least 90% of patients with typical pulmonary valve stenosis). The frontal QRS axis of the electrocardiogram exceeded +170° in five cases. Two patients (cases 3 and 5) had an associated patent foramen ovale.

Echocardiographic imaging was carried out with an ATL Mark 600 or Ultramark 8 mechanical sector scanner ultrasound system (Advanced Technology Laboratories Inc, Bellevue, WA). Images were obtained from the standard and high parasternal short axis views. When we simultaneously measured Doppler echocardiographic gradients and intracardiac pressures in 39 patients with pulmonary valve stenosis we obtained a good correlation (r = 0.95) (gradient [catheter] = 0.89 [gradient [echo]] + 3.98).

Angiocardiography was carried out under ketamine anaesthesia before balloon dilatation. The details of our dilatation procedure have been reported elsewhere.18 Briefly, systemic and right ventricular pressures are continuously monitored before and during dilatation. Balloon catheters with diameters that are 20-30% greater than annulus diameter measured from the lateral right ventriculogram (table 1) are positioned across the pulmonary valve by means of a guide wire in the left pulmonary artery. Inflation-deflation cycles are rapid (less than 15 seconds) and they are repeated (usually for two or three cycles) until no waist can be identified when the balloon is inflated.

Patient 1, in whom pulmonary valve dysplasia had been identified clinically and angiographically, underwent balloon dilatation at the time of surgery. Balloon size was selected according to the preoperative angiogram and the balloon was placed across the pulmonary valve ring from the pulmonary artery and inflated once after cardiopulmonary bypass had been started.

Results

Table 1 shows the results of balloon dilatation. The balloon catheter could not be passed across the pulmonary valve in one patient (case 4). In the operating room, before dilatation, patient 1 was seen to have thickened "fleshy" valve leaflets. A tear was seen in the anterior commissure of this valve as a result of dilatation. This suggests an element of commissural fusion, and in this instance the orifice increased from 3 to 10 mm according to Hegar dilator sizing.

During percutaneous balloon dilatation in the remaining five cases no waist was seen during the initial inflation cycle. Immediately after the procedure the mean pressure gradient across the pulmonary valve decreased insignificantly from 58 (28) to 47 (12) mm Hg. Doppler echocardiography in four patients (cases 2, 3, 5, and 7) six months after operation showed no overall significant relief of stenosis. One
patient (case 5), however, did have a significant reduction in gradient from 60 mm Hg immediately after dilatation to 23 mm Hg six months later (table 1). The pulmonary valve showed features of commissural fusion in addition to dysplasia. On the angiogram the base of one of the leaflets appeared to be thin and a systolic jet was noted (fig 1). An echocardiogram (fig 1) of the pulmonary valve from the long axis view showed doming as well as leaflet thickening, with widening of one of the sinuses of Valsalva in diastole. Thus although the valve leaflets were thickened, there was an element of commissural fusion to account for the success of balloon dilatation.

Table 2 shows the angiographic and echocardiographic features in these patients. All patients showed angiographic evidence of thickening of pulmonary valve leaflets. A supra-annular ridge was noted in all but one patient (case 7) (fig 2). Patients 5 and 7 did not show asymmetric systolic doming, and a systolic jet was seen in four of the seven patients. Post-stenotic dilatation of the proximal main pulmonary artery was present in two patients (cases 4 and 7). Patients 5 and 7 showed diastolic widening of one sinus of Valsalva on the lateral projection of the right ventriculogram, suggesting that in addition to dysplasia there was commissural fusion between at least two adjacent cusps. None of the patients had a fixed subvalve pulmonary stenosis. The mean (SD) diameter of the pulmonary valve annulus was 10-7 (2-11) mm and this is 90 (18) % of the predicted normal for body weight.19

Table 2  A comparison of angiographic and echocardiographic features in seven patients with dysplastic pulmonary valves

<table>
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<tr>
<th>Patient No:</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
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<th>6</th>
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<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Thickened leaflets</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Asymmetrical systolic doming</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Supra-annular narrowing</td>
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<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
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<tr>
<td>Systolic jet</td>
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<td>o</td>
<td>o</td>
<td>+</td>
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<td>+</td>
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<td>Thickened leaflets</td>
<td>+</td>
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<td>+</td>
<td>+</td>
<td>+</td>
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<td>+</td>
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<td>Immobility of leaflets</td>
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<td>+</td>
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<tr>
<td>Supra-annular narrowing</td>
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<td>+</td>
<td>+</td>
<td>+</td>
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<td>o</td>
<td>o</td>
<td>+</td>
<td>o</td>
<td>o</td>
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</tr>
</tbody>
</table>

+, present; o, absent.
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Fig 2  (a) Echocardiogram of precordial long axis view from a patient with a dysplastic pulmonary valve. Note the bright echoes originating from the thickened valve. There is also evidence of supravalvar narrowing (black arrows). (b) Angiocardiogram in the same patient. Note the absence of a central jet, the supravalvar narrowing (black arrows), and a fleshy appearance of the valve leaflets. (c) Short axis echocardiographic view. (d) Photograph of long axis cut from another patient with a dysplastic pulmonary valve showing fleshy and thickened valve leaflets. There is no evidence of fusion along the commissures. ac, anterior cusp; lc, left cusp; rc, right cusp; mpa, main pulmonary artery; pv, pulmonary valve; rv, right ventricle.

The cross sectional echocardiographic features that were common to patients with dysplasia correlated closely with those seen during ventriculography (figs 1–2 and table 2). The high parasternal short axis view revealed tricuspid pulmonary valves in all patients. Both the shape and the mobility of individual valve leaflets could be determined by combining the parasternal long and short axis views;
all patients had thickened leaflets. Poor leaflet mobility could be seen in both long and short axis views in all patients except cases 5 and 7. The anterior sinus of Valsalva in each of these patients showed diastolic widening in the parasternal long axis view—a feature not seen in either the posterior sinuses of Valsalva in these two cases or in any of the other patients. Post-stenotic dilatation of the main pulmonary artery was seen in one patient only (case 7).

Discussion

The association of a particular phenotype with pulmonary valve stenosis has been well recognised since the original description by Noonan in the early 1960s, with up to 50% of patients having dysplasia of the valve as the mechanism of stenosis. It is very likely that the wide variation seen in the physical features of this syndrome is similarly expressed in variable degrees of valve dysplasia. Other clinical...
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The dysplastic pulmonary valve stenosis is a form of valvular pulmonary stenosis characterized by a dysplastic or malformed pulmonary valve. It is often associated with other cardiac anomalies, such as hypoplastic left heart syndrome, tetralogy of Fallot, and other congenital heart defects. The valve may be thickened, malformed, or have reduced leaflet mobility due to fusion or dysplasia.

Feasibility of diagnosis and treatment

The presence of a dysplastic pulmonary valve can be suspected on clinical examination, echocardiography, and cardiac catheterization. Echocardiography is particularly useful in identifying the degree of stenosis, the presence of a jet, and the presence of other associated congenital heart defects.

Treatment

Treatment options for dysplastic pulmonary valve stenosis include balloon valvuloplasty, surgical valve replacement, or valve repair. The choice of treatment depends on the severity of the stenosis, the presence of other cardiac anomalies, and the patient's overall medical condition.

References


