Balloon Dilation Angioplasty of Hypoplastic and Stenotic Pulmonary Arteries

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SUMMARY Balloon dilation angioplasty (BDA) was attempted in seven children with either stenosis or hypoplasia (a narrowing greater than 1 cm long extending past the lung hilum) of both right and left pulmonary arteries (PAs). In two of these seven, the procedure could not be performed because of technical difficulties. In each of the remaining five children (1½–16 years old), the right ventricular (RV) pressure was greater than ½ left ventricular pressure, main PA pressure was greater than 60 mm Hg, and previous operative attempts to relieve RV outflow obstruction, including the branch PA obstruction, were unsuccessful. BDA was performed in only one PA in each patient, and was considered successful if the pressure gradient decreased, angiographic diameter increased, and the percentage of blood flow increased from the dilated lung. BDA was successful in all five children; RV pressure fell from 104 ± 42 to 80 ± 30 mm Hg (p < 0.05), the gradient across the obstruction fell from 61 ± 51 to 32 ± 22 mm Hg (p < 0.05), the diameter of the narrowed segment increased from 3.7 ± 1.2 to 6.8 ± 1.1 mm, p = 0.02, and the percentage of blood flow (as determined by quantitative lung scan) to the dilated lung increased from 41 ± 16% to 52 ± 22% (p < 0.05). No morbidity was observed in any patient. Follow-up angiograms (2–12 months) in three of five patients indicate persistence of the anatomic improvement. While BDA did not restore right-heart pressures and anatomy to normal, it provided significant hemodynamic relief to a group of patients in whom traditional operative management has usually been unsuccessful. Final determination of the role of BDA in such patients must await the results of further studies.

THE advent of a safe and effective method for dilating atherosclerotic lesions in peripheral arteries1,2 promptly led to the extension of this approach to coronary arteries,3 renal artery stenosis,4 and other lesions throughout the body. The results of balloon dilation angioplasty in these acquired arteriopathies have, in general, been gratifying. However, the extension of balloon dilation angioplasty to children with congenital vascular and cardiac narrowings has been slow. There are good reasons for caution. Congenital stenoses have no soft intimal plaque that may be "squeezed" by the balloon; the goal of therapy is not to restore a vessel lumen, but to enlarge an abnormally small structure; congenital lesions require treatment during growth; and if dilatation produces vessel scarring, vascular growth may be impaired. Also, since most important congenital stenoses involve cardiac valves or large vessels near the heart, the ability of an already compromised circulation to tolerate prolonged occlusion of such a vessel is not established. As a result, the current clinical literature on dilating congenital lesions consists of isolated case reports3–7 or small series8 of cases, with variable results.

Postmortem9, excised specimen10 and animal studies11–14 indicate that balloon dilatation of congenital cardiac defects may, under certain circumstances, be safe and effective. Armed with guidelines and experience derived from such studies, we instituted a clinical trial of balloon dilation angioplasty in congenital cardiac lesions in January 1981. Initially, we chose to dilate a lesion for which traditional surgical approaches have been particularly unrewarding: the hypoplastic or stenotic pulmonary artery.

Materials and Methods

Patients

Before considering a child for balloon dilation, several hemodynamic and angiographic findings were sought: congenital stenosis or hypoplasia of one or more branch pulmonary arteries; a diameter of the narrowed segment of 6 mm or less; right ventricular pressure greater than ½ left ventricular pressure without associated left-to-right shunt; main pulmonary artery (MPA) pressure of 60 mm Hg or greater; and previous operative attempt at relief of right ventricular outflow obstruction. Children who met all of the above requirements were presented at the combined Pediatric Cardiology/Surgery/Radiology conference at the University of Minnesota. If the conference consensus was that further attempts at gradient relief were warranted, the parents of those children were offered either an attempt at experimental dilation of the pulmonary artery or traditional surgical management. If the pulmonary arterial narrowing was greater than 1 cm long and extended past the hilum of the lung, the parents were offered either dilation angioplasty or medical manage-
ment. Informed consent from the parents (and the child, when appropriate) was obtained.

Assessment of the Effects of Dilation Angioplasty

Successful relief of a vascular obstruction should increase the diameter of the obstructed segment, decrease the pressure gradient and increase blood flow across the site. The diameter of the pulmonary artery was measured directly from biplane cineangiograms; magnification was corrected for using the known catheter diameter. Pressure gradients were measured with fluid-filled catheters connected to Statham 23Db strain gauges (zeroed to midchest level) and recorded on an E for M VR 12 recorder. The proportion of total pulmonary blood flow directed to the dilated lung was determined by quantitative lung scans. Technetium-99 tagged to macroaggregated albumin was injected into a peripheral vein and the total number of counts in each lung was measured with a CE maxicamera II gamma camera with a large field of view. The quantitative lung scan in patient 1 was invalid for technical reasons; otherwise, each variable was measured before and after dilation in each patient.

Protocol for Dilation Angioplasty

Our experience in lambs was used to design the initial dilation protocol.11 In lambs, balloons 2–2½ times the size of the narrowed pulmonary artery would evidence a “waist” when inflated to 1 atmosphere; that waist disappeared at an inflation pressure of 4–5 atmospheres, resulting in invariable evidence of intimal tears at the dilated site. Therefore, we did not advance an unguided catheter or wire across a previously dilated site. In each patient, we advanced an endhole catheter across the narrowed site and threaded a 0.038-inch Teflon-coated guidewire through the catheter. The introducing catheter was exchanged for a polyethylene balloon dilation catheter (Meditech, Inc. or Cook, Inc.). The balloon dimensions of the dilating catheter (usually #8F) depended on the estimated diameter of the lesion to be dilated. After positioning in the distal pulmonary artery tree, the balloon was inflated (with 60% diatrizoate meglumine diluted 1:1 with normal saline) to 1 atmosphere and held at that pressure level (as determined by a strain gauge). If no waist was seen in the balloon, the balloon was withdrawn slowly through the pulmonary artery until a waist was seen on biplane fluoroscopy. If no waist was seen at any time, the dilating catheter was removed and replaced by a catheter whose balloon diameter was 2 mm larger than that previously used.

Once a waist was visualized near the middle of the balloon, half-strength contrast was delivered into the balloon until the waist disappeared or until a dilating pressure of 7–8 atmospheres was achieved. Pressure was held in the balloon for at least 30 seconds; arterial blood pressure was monitored by an indwelling line. After dilation, a #7F pigtail angiographic catheter was advanced over the guidewire after removing the dilating catheter, and a postdilation angiogram was performed.

If a clear-cut waist was not visible at 1 atmosphere using the largest available balloon, that balloon was nonetheless positioned directly underneath the stenotic area and inflated to 7–8 atmospheres for 1 minute. In the last two patients, if the postdilation angiogram did not demonstrate at least a twofold increase in the diameter of the hypoplastic/stenotic segment, the balloon diameter was increased by 2 mm and a final dilation was performed.

All dilations were performed in the cardiac catheterization laboratory. Each patient was presedated with securbarital (2 mg/kg) and morphine (0.1 mg/kg) intramuscularly. Additional sedation was administered as required with either diazepam or additional morphine. Venous access was obtained percutaneously via the right femoral vein in each patient. At least two units of blood were available in the catheterization laboratory, and cardiovascular surgical standby was arranged for each child. After dilation, the chest was fluoroscoped for signs of extravascular bleeding, and each patient was monitored overnight in the pediatric intensive care unit. General anesthesia was not used in any patient.

Results

Patients

Seven children were enrolled in this protocol. In one child, status post tetralogy of Fallot repair with markedly diminished flow to the left lung, all attempts to pass a catheter into the left pulmonary artery (LPA) were unsuccessful. At subsequent operation, the orifice to the LPA was found to be 1 mm. A second child, also with tetralogy of Fallot status post complete repair, the LPA arose from the right pulmonary artery (RPA) at an acute angle of 40°, opposite to the blood flow. Although multiple catheters and guidewires were advanced across the narrowed segment, we could not get a stiff dilating catheter to follow the wire.

The ages, diagnoses and involved artery of the five children whose pulmonary arteries were successfully dilated are listed in table I. One child underwent dila-

<table>
<thead>
<tr>
<th>Pt</th>
<th>Age (years)</th>
<th>Cardiac lesion</th>
<th>Artery dilated</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>14</td>
<td>Tetralogy of Fallot status postrepair</td>
<td>LPA</td>
</tr>
<tr>
<td>2</td>
<td>16</td>
<td>Tetralogy of Fallot status postrepair</td>
<td>LPA</td>
</tr>
<tr>
<td>3</td>
<td>8</td>
<td>Tetralogy of Fallot status postrepair</td>
<td>LPA</td>
</tr>
<tr>
<td>4</td>
<td>1½</td>
<td>VSD, hypoplastic pulmonary arteries</td>
<td>RPA</td>
</tr>
<tr>
<td>5a</td>
<td>7</td>
<td>Tetralogy of Fallot, hypoplastic pulmonary arteries</td>
<td>LPA</td>
</tr>
<tr>
<td>5b</td>
<td>7½</td>
<td>Tetralogy of Fallot, hypoplastic pulmonary arteries</td>
<td>RPA</td>
</tr>
</tbody>
</table>

*Patient 5 underwent balloon dilation twice.

Abbreviations: VSD = ventricular septal defect; LPA = left pulmonary artery; RPA = right pulmonary artery.
tion on two separate occasions. In all children, both the RPA and LPA were either stenotic or hypoplastic, producing right ventricular hypertension rather than just a flow distribution abnormality. In patients 1–3, the LPA stenosis was more severe than that of the RPA (both angiographically and by flow distribution), so the LPA was preferentially dilated. In patients 4 and 5, with hypoplastic pulmonary arteries, the arteries to each lung were equally affected.

Protocol for Dilation Angioplasty

In the first three children, the LPA diameters were 4.0 mm or larger. In each, inflation of an 8- or 10-mm balloon to 1 atmosphere (which, in animal studies is too low a pressure to produce intimal tears) did not induce a waist in the balloon. Thus, human congenital branch pulmonary arterial stenosis is an extremely compliant lesion that can stretch to more than twice its resting diameter without apparent intimal tears. Such a dilation produced no increase in diameter on postdilation angiograms. In those children, we therefore used the largest balloons then available (before December 1981) — 10–12 mm. In those three children, the effects of dilation were relatively modest (table 2). As a result of this experience, we obtained and used progressively larger balloons, starting with diameters three times the diameter of the narrowed segment.

The dilation procedures in these five children took an average of 160 minutes, with an average of seven catheter changes, and required an average of 48 minutes of fluoroscopy time.

Effects of Dilation Angioplasty

Dilation angioplasty was successful in all five patients to varying degrees. In each, angioplasty decreased the gradient (p < 0.05), increased the diameter (p < 0.02), and increased the flow (p < 0.05) at the narrowed site (table 2). When the dilating balloon was less than three times the diameter of the narrowed site, the diameter changes (fig. 1) and hemodynamic effects of dilation were modest; when the diameter of the dilating balloon approached four times the diameter of

### Table 2. Hemodynamic and Angiographic Summary of Balloon Dilation

<table>
<thead>
<tr>
<th>Pt</th>
<th>Diameter of dilating balloon (mm)</th>
<th>RV pressure (mm Hg)</th>
<th>PA diameter (mm)</th>
<th>% flow to lung</th>
<th>Branch PA-to-MPA gradient</th>
<th>RV pressure (mm Hg)</th>
<th>PA diameter (mm)</th>
<th>% flow to lung</th>
<th>Branch PA-to-MPA gradient</th>
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<tr>
<td>1</td>
<td>9</td>
<td>74</td>
<td>5.5</td>
<td>—</td>
<td>22</td>
<td>56</td>
<td>7.4</td>
<td>—</td>
<td>15</td>
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<tr>
<td>2</td>
<td>12</td>
<td>79</td>
<td>4.5</td>
<td>25</td>
<td>35</td>
<td>68</td>
<td>6.6</td>
<td>30</td>
<td>29</td>
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<tr>
<td>3</td>
<td>12</td>
<td>72</td>
<td>4.1</td>
<td>30</td>
<td>30</td>
<td>52</td>
<td>6.0</td>
<td>36</td>
<td>15</td>
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<td>55</td>
<td>145</td>
<td>119</td>
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<tr>
<td>5†</td>
<td>a</td>
<td>10</td>
<td>2.7</td>
<td>55</td>
<td>75</td>
<td>107</td>
<td>6.0</td>
<td>67</td>
<td>35</td>
</tr>
<tr>
<td>b</td>
<td>112</td>
<td>2.2</td>
<td>27</td>
<td>25</td>
<td>5</td>
<td>101</td>
<td>6.8</td>
<td>39</td>
<td>5</td>
</tr>
<tr>
<td>Mean</td>
<td>±SD</td>
<td>105.3</td>
<td>3.5</td>
<td>38.4</td>
<td>55.3</td>
<td>83.8*</td>
<td>6.8*</td>
<td>49.2*</td>
<td>27.8*</td>
</tr>
</tbody>
</table>

*p < 0.05 compared with predilation values, paired t test.
†Patient 5 underwent balloon dilation twice.
Abbreviations: RV = right ventricular; PA = pulmonary artery; MPA = main pulmonary artery.

Figure 1. Pre- and postdilation angiograms of the least successful dilation (patient 2). The proximal left pulmonary artery is seen on a lateral projection; the arrows indicate the narrowest portion of the vessel. The irregularities may indicate intimal tears in the postdilation angiogram.
the narrowed pulmonary artery (patients 4 and 5), both the angiographic (figs. 2 and 3) and the hemodynamic effects (table 2) of dilation were more impressive.

There were further signs of improvement after dilation. In patient 4, the distal RPA oxygen saturation was higher than that of the MPA (70% vs 63%) before dilation, suggesting a significant systemic arterial contribution to distal pulmonary arterial blood flow. After dilation, RPA and MPA saturations were identical (65%), and retrograde filling of a systemic-pulmonary collateral artery was visible. In patient 5, who had a nonrestrictive ventricular septal defect, an average of four aortic oxygen saturations before dilation (74%) rose after dilation (86%), and evidence of right-to-left shunting on the lung scan diminished (fig. 4).

The fall in gradient in patients with isolated branch pulmonary arterial stenosis was due to a decrease in MPA pressure alone; distal pressures remained normal (less than 30 mm Hg systolic) after dilation. In the two children with hypoplastic pulmonary arteries, distal pressure after dilation was elevated (51 ± 13 and 36 ± 12 mm Hg), suggesting the presence of residual stenosis beyond the hilum of the lung in those children.

Dilation angioplasty was well tolerated in all five

![Figure 2. Pre- and postdilation angiograms in patient 4. The entire central right pulmonary artery is hypoplastic, with the narrowings extending into the hilum of the lung, past the lobar branch points. Two areas of particularly severe stenosis (arrows) may be seen. After dilation, the entire main right pulmonary artery has enlarged, with an improved appearance of the right hilum. There is a definite irregularity at the takeoff of the right pulmonary artery (arrow), which presumably represents an intimal tear. The hypoplastic left pulmonary artery is faintly visible, as is a surgically related false aneurysm arising from the main pulmonary artery.](http://circ.ahajournals.org/)

![Figure 3. Pre- and postdilation angiograms from patient 5. A lateral view of the proximal left pulmonary artery demonstrated a stenosis (arrow) extending into the hilum of the left lung and involving the origin of the left lower and upper lobe vessels. After dilation, improved angiographic appearance is associated with a clear-cut irregularity on the inferior margin of the dilated segments (arrow), presumably due to an intimal tear.](http://circ.ahajournals.org/)
children: there were no changes in chest x-ray, vital signs, or hemoglobin levels to suggest significant blood loss. Each patient was monitored for 24 hours in the pediatric intensive care unit and none had evidence of hemodynamic instability. The patients were discharged 36–60 hours after dilation. There was, however, clear evidence on the angiograms of irregularities of the vascular intima after dilation at the site of obstruction in several patients (figs. 2 and 3), presumably resulting from intimal tears induced by the dilatations. These tears appeared well tolerated (although the children complained of chest pain at the moment of dilation).

Late Follow-up

Each child has remained well and without signs of subsequent deterioration during a follow-up of 2–15 months. Three (patients 1, 4 and 5) have undergone repeat cardiac catheterization; in each, the diameters of the narrowed site remained similar to their postdilation values (figs. 2 and 5), and follow-up perfusion lung scans in patients 4 and 5 indicated a further small increase in the proportion of flow directed to the dilated lung.

In patient 5, the hemoglobin level was above 21 g% before dilation, but has remained below 16 g% since dilation, with a concomitant decrease in cyanosis.

Discussion

Branch pulmonary arterial stenosis and hypoplasia can occur as isolated lesions, although more often they complicate tetralogy of Fallot or tetralogy of Fallot with pulmonary atresia. Despite the recent advances in congenital heart surgery, reconstruction of a stenotic or hypoplastic pulmonary artery remains a difficult problem. Thus, despite the well-known association between branch pulmonary arterial stenosis and poor results after tetralogy of Fallot surgery, operative mortality has remained high (and residual stenosis common) in that group of patients. In the specific instance of RPA stenosis after Waterston anastomoses, dismantling of the shunt and reconstruction of the RPA is associated with a significant incidence of residual obstruction.

If operative relief of branch pulmonary arterial obstruction remains problematic, operative management of hypoplastic pulmonary arteries has been frankly disappointing. Attempts to promote pulmonary arterial growth with systemic-to-pulmonary artery or right ventricular-to-pulmonary artery anastomoses have been the bulwark of therapy. This approach may increase the size of the central pulmonary artery, although the evidence that such maneuvers actually increase blood flow to the affected lung is unconvincing.

Within this context, the use of balloon dilation angioplasty to enlarge stenotic or hypoplastic pulmonary arteries appears to be a reasonable therapeutic option. Despite the small number of patients reported here, the results and risk of the procedure do not appear worse than those of operative series. More important, dilation angioplasty appears to increase not only the size of, but also the blood flow through, hypoplastic arteries. Increasing blood flow through such vessels is not likely to result from a traditional operative approach. Haworth et al. noted that direct surgical anastomosis into a hypoplastic pulmonary artery generally uncovered stenosis distal to the mediastinum. McGoon et al. noted that such distal stenoses are usually confined to the origins of the lobar arteries, a region of the lung within reach of a dilating balloon (fig. 2). Thus, the use of balloon dilation angioplasty to address sten-
sues of both the first and second generation of pulmonary arterial branches appears to offer the best chance of salvaging hypoplastic pulmonary arteries. Since the intraacinar arteries of children with congenital pulmonary arterial stenosis are decreased in number,21 and since the growth and development of such vessels appears to be greatest in the first 2 years of life,22 we anticipate that successful dilations performed before the age of 2 years might yield the best long-term results. The hypothesis that the distal pulmonary circulation in such patients is indeed abnormal is supported by the observation in patients 4 and 5 that distal pulmonary arterial pressure became elevated after dilation.

In many ways, balloon dilation of pulmonary arterial stenoses reflects the experience in animals.11 Human experience does differ in one very important respect: Human congenital pulmonary arterial stenosis is a very compliant lesion, stretching to over twice its native diameter without signs of intimal tears. Much larger balloons appear to be required to tear the intima of pulmonary arterial lesions. The optimal balloon size for a given child must therefore be established at the time of dilation.

Although no significant morbidity was encountered in these patients undergoing balloon dilation angioplasty, several short- and long-term potential complications can be identified: vascular rupture from an overlarge balloon, extravascular passage of a guidewire through an intimal tear, protrusion of the balloon into the MPA with concomitant complete obstruction of right ventricular outflow, and the late development of aneurysms and accelerated atherosclerosis. In addition, despite the encouraging results of this initial approach to dilation of hypoplastic or stenotic pulmonary arteries, a number of factors may limit its clinical value. Several investigators have reported encouraging initial results with clear-cut clinical or hemodynamic deterioration within 2 months.8,25 Much longer follow-up is required in our patients before balloon dilation angioplasty of stenotic and hypoplastic pulmonary arteries assumes a primary role in the management of congenital cardiac disease.

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Circulation. 1983;67:962-967
doi: 10.1161/01.CIR.67.5.962

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
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