Balloon Valvotomy for Critical Stenosis or Atresia of Pulmonary Valve in Newborns

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Objectives. Percutaneous balloon valvotomy was studied retrospectively in newborns with critical pulmonary valve stenosis or atresia to assess its potential role as an alternative therapy to operation.

Background. Severe right ventricular outflow tract obstructions are life-threatening conditions requiring prostaglandin infusion immediately after birth and then relief of the valvular obstruction. To avoid surgical hazards at this age, it would be useful to extend to newborns the balloon valvotomy so effective in older patients.

Methods. Ninety-seven newborns (82 with critical pulmonary valve stenosis, 15 with atresia) underwent balloon valvotomy, provided that they had a well-developed right ventricle, including an infundibulum close to the pulmonary artery. In patients with atresia, the outflow tract membrane had to be perforated with a wire needle or a radiofrequency probe.

Results. Balloon valvotomy could be performed in 81 patients and was effective in 77. It caused 3 fatal and 16 non-fatal complications. Ten patients with persistent poor right ventricular compliance despite an effective valvotomy required a surgical shunt. Among the 81 patients in whom the procedure could be performed, right ventricular surgery was avoided in 5 (55%) of the 9 patients with atresia (95% confidence interval [CI] 28% to 80%) and 55 (76%) of the 72 patients with stenosis (95% CI 66% to 86%) at the end of the follow-up period (9.7 years).

Conclusions. Balloon pulmonary valvotomy is not always feasible in newborns, but it is relatively safe and effective and should be considered a valid alternative to operation.

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Since it was first performed in 1982 (1), percutaneous balloon valvotomy has become the elective treatment of pulmonary valve stenosis in children. However, in newborn infants, critical pulmonary valve stenosis raises specific problems because of frequent association with hypoplasia of the right ventricle. Right ventricular hypoplasia results in decreased right ventricular compliance and right to left atrial shunting, causing cyanosis. The physiology of pulmonary valve atresia, in which right ventricular hypoplasia is even more severe, is very similar. Both conditions are emergent and require prompt prostaglandin therapy to keep the ductus arteriosus open and then relief of the obstruction so that blood can flow into the pulmonary artery through the pulmonary valve. For a long time, operation was the only way to relieve the obstruction (2,3). The only published reports of balloon valvotomy in these patients were preliminary studies only dealing with a limited number of cases (4-9). The present retrospective study sought to assess whether percutaneous balloon valvotomy is a valid alternative therapy to operation. We therefore report our experience with attempted dilation or perforation and dilation of the obstructed valve in 97 consecutive newborns (82 with pulmonary valve stenosis, 15 with pulmonary valve atresia) in our catheterization laboratory. To our knowledge, this is the largest series of pulmonary balloon valvotomies performed within the first month of life.

Methods

Patients. All patients admitted from September 1984 to March 1994 with critical pulmonary valve stenosis were included in this retrospective study. After April 1990, eligible newborns with pulmonary valve atresia were also included. Demographic data are shown in Table 1.

Diagnoses were made by two-dimensional echocardiography and Doppler analysis (10). We could thus obtain direct measurements of the size of the pulmonary valve annulus as well as Doppler estimation of right ventricular systolic pressure and the systolic pressure gradient across the pulmonary valve.

All patients had a tripartite right ventricle with a tricuspid annulus size ranging from 8 to 15 mm (mean ±SD 11.5 ± 1.6). All right ventricles had a well-developed outflow tract in contact with a normal-sized main pulmonary artery. The obstruction consisted of a thick dome in patients with pulmonary valve stenosis or an imperforate membrane in those with pulmonary valve atresia. During this study, 31 patients with pulmonary valve atresia and extreme right ventricular hypoplasia...
Pulmonary Valve surgery, which we called right ventricular valvotomy, was undertaken in patients under local anesthesia using the pulmonary valve and its inflation; 2) pressure equal to or greater than systemic pressure. They also all had a right to left shunt through the foramen ovale, leading to systemic oxygen saturation <90%. A lateral right ventricular catheterogram was obtained. In the 82 patients with pulmonary valve stenosis, the main pulmonary artery was filled by a thin jet of contrast medium crossing a thick valvular “dome” with a small orifice (Fig. 1A) or by a “cloud of smoke” that barely allowed clear visualization of the main pulmonary artery (Fig. 1B). This “cloud of smoke” was very difficult to see on the cineangiogram but easier on a digitalized angiogram (Fig. 1B). In contrast, in the 15 patients with pulmonary valve atresia, no medium at all crossed a “dead-ended” right ventricular outflow tract (Fig. 1C). None of these patients had significant coronary-right ventricular sinusoidal connections. A left ventriculogram was then obtained to check ventricular septal integrity and to confirm that the pulmonary artery injected through the ductus arteriosus was in contact with the infundibulum.

After standard catheterization, balloon valvotomy was performed, with the type of obstruction (pulmonary valve stenosis or atresia) determining the technique used. In most patients with pulmonary valve stenosis, the pulmonary valve was first crossed by an end-hole catheter to record pulmonary artery pressures. In 12 patients with pulmonary valve stenosis with a very small valvular orifice, the pulmonary valve was first crossed with a wire balloon for coronary angioplasty (Probe Bard) driven in a 5F Right Judkins catheter (Cordis) to predilate it and then with an end-hole catheter. A steerable guide wire was then passed into this catheter and advanced as far as possible into the left pulmonary artery. We avoided advancing the guide wire into the descending aorta through the ductus arteriosus, which may close suddenly in reaction to endothelial damage. In fact, sudden closure of the ductus arteriosus occurred early in our experience, requiring urgent operation.

The balloon dilation catheter was chosen to match a balloon-annulus diameter ratio of 110% to 190% (mean 136 ± 48%). Balloon size thus ranged from 8 to 15 mm. We tried many types of balloon, including Trefoil Schneider (3 × 5 to 3 × 7 mm) and PTA 5F Bard, and later in the study we used the TyShak balloon (NuMed). After introducing the balloon over the wire and appropriately positioning it across the pulmonary valve, we inflated the balloon manually once or twice until the thin valve waist disappeared. However, we

### Table 1. Demographic Data for 97 Study Patients

<table>
<thead>
<tr>
<th></th>
<th>Pulmonary Valve Stenosis (n = 82)</th>
<th>Pulmonary Valve Atresia (n = 15)</th>
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</thead>
<tbody>
<tr>
<td>Mean age (days)</td>
<td>8.06</td>
<td>3.78</td>
</tr>
<tr>
<td>Mean weight (kg)</td>
<td>3.17</td>
<td>2.90</td>
</tr>
<tr>
<td>Cyanosis (O2 sat &lt;90%)</td>
<td>82 (100%)</td>
<td>15 (100%)</td>
</tr>
<tr>
<td>Deep cyanosis (O2 sat &lt;80%)</td>
<td>54 (66%)</td>
<td>15 (100%)</td>
</tr>
<tr>
<td>Urgent need of IV PGE1</td>
<td>42 (51%)</td>
<td>15 (100%)</td>
</tr>
</tbody>
</table>

Unless otherwise indicated, data presented are number (%) of patients. IV PGE1 = intravenous infusion of prostaglandin E1, O2 sat = oxygen saturation.

**Study design.** To determine whether balloon valvotomy is a valid alternative to conventional operation for critical pulmonary valve stenosis and pulmonary valve atresia in newborns, we retrospectively analyzed data from 97 balloon valvotomies performed in 82 infants with pulmonary valve stenosis and 15 with pulmonary valve atresia. We assessed feasibility, safety, effectiveness and ultimate success. We considered balloon valvotomy 1) feasible if the whole procedure could be performed until placement of the balloon across the pulmonary valve and its inflation; 2) safe if the infant survived and did not have serious complications; 3) effective if it relieved outflow obstruction, as indicated by a systolic pressure gradient across the valve ≤40 mm Hg, a right ventricular systolic pressure ≤60 mm Hg and a right ventricular/aortic systolic pressure ratio equal ≤0.8; and 4) ultimately successful if any pulmonary valve surgery, which we called right ventricular surgery, was avoided, even if we had to place a surgical shunt temporarily because of persistent poor right ventricular compliance.

**Methods.** All patients underwent standard catheterization before the balloon valvotomy itself. Right heart catheterization was undertaken in patients under local anesthesia using the femoral venous approach. Pressures were recorded and oxygen saturations measured in all heart chambers through the patent foramen ovale. All patients had a right ventricular systolic pressure equal to or greater than systemic pressure. They also all had a right to left shunt through the foramen ovale, leading to systemic oxygen saturation <90%. A lateral right ventricular catheterogram was obtained. In the 82 patients with pulmonary valve stenosis, the main pulmonary artery was filled by a thin jet of contrast medium crossing a thick valvular “dome” with a small orifice (Fig. 1A) or by a “cloud of smoke” that barely allowed clear visualization of the main pulmonary artery (Fig. 1B). This “cloud of smoke” was very difficult to see on the cineangiogram but easier on a digitalized angiogram (Fig. 1B). In contrast, in the 15 patients with pulmonary valve atresia, no medium at all crossed a “dead-ended” right ventricular outflow tract (Fig. 1C). None of these patients had significant coronary-right ventricular sinusoidal connections. A left ventriculogram was then obtained to check ventricular septal integrity and to confirm that the pulmonary artery injected through the ductus arteriosus was in contact with the infundibulum.

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**Figure 1. Morphology of right ventricular outflow tract in newborns with pulmonary valve stenosis or atresia.** Right ventricular digitized angiograms in the lateral projection. In all infants, the right ventricle is well developed, including a normal outflow chamber. A, Critical pulmonary valve stenosis. A thin jet of contrast medium crosses a thick but open dome-shaped valve and fills a dilated pulmonary artery (P). B, Severe critical pulmonary valve stenosis. A “cloud of smoke” (arrow) passes through a pinhole in the valve. C, Pulmonary valve atresia. The outflow tract is “dead-ended,” with no medium at all passing through it (asterisk).
Table 2. Early Outcome* of Balloon Valvotomy in Newborns

<table>
<thead>
<tr>
<th></th>
<th>Pulmonary Valve Stenosis (n = 82)</th>
<th>Pulmonary Valve Atresia (n = 15)</th>
<th>Total (n = 97)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Procedural failure</td>
<td>10</td>
<td>6</td>
<td>16</td>
</tr>
<tr>
<td>Procedural success</td>
<td>72</td>
<td>9</td>
<td>81</td>
</tr>
<tr>
<td>Death</td>
<td>2</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>Ineffective</td>
<td>2</td>
<td>0</td>
<td>2</td>
</tr>
<tr>
<td>Effective</td>
<td>68</td>
<td>9</td>
<td>77</td>
</tr>
<tr>
<td>Shunt required</td>
<td>5</td>
<td>5</td>
<td>10</td>
</tr>
<tr>
<td>Shunt not required</td>
<td>63</td>
<td>4</td>
<td>67</td>
</tr>
</tbody>
</table>

*Within 1 month of balloon valvotomy. †Two patients later died, one from persistent atrial flutter induced by the attempt at balloon valvotomy and the other after operation for pulmonary valve atresia.

Results

Critical pulmonary valve stenosis. Early results. Of 82 patients with critical pulmonary stenosis, balloon valvotomy could be performed in 72 but not (failed procedures) in 10 (12%, 95% confidence interval [CI] 4% to 20%) (Table 2). However, there were only two failures after 1990. The reasons for the 10 failures were inability to cross the valve in eight patients and interruption of the procedure because of a complication in two. Of these 10 patients, 9 underwent right ventricular surgery within the next few days; 1 of the 9 died postoperatively. One patient did not undergo operation because he developed severe necrotizing enterocolitis; however, his condition ultimately improved well enough for operation at 21 months of age.

In the 72 patients (88%, CI 80% to 96%) with balloon valvotomy, right ventricular systolic pressure decreased by 50% (from 114 ± 22 mm Hg [range 80 to 180] to 55 ± 17 mm Hg [range 30 to 130], p < 0.001; the systolic pressure gradient across the pulmonary valve decreased by 50% (from 84 ± 27 mm Hg [range 30 to 166] to 36 ± 11 mm Hg [range 30 to 60], p < 0.001); and the right ventricular/aortic systolic pres-
pressure ratio decreased by 35% (from 1.13 ± 0.3 [range 0.8 to 2.18] to 0.73 ± 0.26 [range 0.35 to 1.54], p < 0.001).

Two patients died shortly after balloon valvotomy. One of the two died after subsequent endomyocardial biopsy for Noonan syndrome with cardiomyopathy. There was no ultrasonic evidence for hemopericardium, but autopsy was not permitted. The second patient died suddenly 6 h after balloon valvotomy that had been complicated by myocardial dissection during angiography.

Two other patients had an ineffective valvotomy, indicated by a residual gradient >50 mm Hg, and underwent successful right ventricular surgery. Sixty-eight patients had an effective valvotomy. Although five of them initially had a high residual gradient, the final outcome was good. The residual gradient was clearly due to a dynamic infundibular reaction because this gradient decreased quickly. In the remaining 63 patients, the procedure was considered an immediate success, as indicated by a low residual gradient (13 ± 12 mm Hg, range 0 to 40).

Fifteen of the 68 patients with an effective valvotomy had a persistent right to left atrial shunt and remained cyanotic despite an unobstructed right ventricle. The first five patients who remained cyanotic underwent a surgical shunt. In contrast, the last 10 patients who remained cyanotic despite an effective valvotomy continued to receive prostaglandin infusion until right ventricular compliance improved. We waited for improvement for a maximum of 22 days (mean 6.6) until oxygen saturation increased to a minimum level of 80%.

Complications. In addition to the 2 aforementioned deaths, there were 12 nonfatal but significant complications related to the procedure (13%, CI 5% to 21%): myocardial damage with hemopericardium, which was drained in the catheterization laboratory; burst of the balloon, which was lost in the inferior vena cava without any consequences; seizures after the procedure with no late sequelae; ventricular tachycardia successfully treated with amiodarone; abrupt closure of the ductus arteriosus in three patients (possibly from ductal “irritation” by wires and catheters; prostaglandins failed to maintain ductus arteriosus patency, and urgent palliative operation was required in one patient); necrotizing enterocolitis leading to several months of intravenous nutrition in two patients; septic shock; and stripping of the iliac vein when the balloon catheter was pulled out in two patients.

Follow-up. Of the 68 newborns surviving an effective pulmonary valvotomy, 2 were lost to follow-up. The remaining 66 patients were followed up for a mean of 26.4 ± 21.9 months (range 0.1 to 9.7 years) (Fig. 3).

The procedure was considered an ultimate success in 55 of these 66 patients, as indicated by a Doppler right ventricular outflow gradient <35 mm Hg. On the basis of the 72 patients with successful balloon valvotomy, the ultimate success rate was 76% (CI 66% to 80%). Of the 55 patients with an ultimate success, 50 were in stable condition after valvotomy and never required either repeat balloon valvotomy or operation; 3 required repeat balloon valvotomy because of restenosis; and 2 initially had poor right ventricular compliance and thus required a surgical shunt. The shunt was no longer needed after a few months because right ventricular compliance improved; it was removed surgically in one patient and clogged spontaneously in the other.

Of 11 patients in whom balloon valvotomy failed to alleviate pulmonary valve stenosis, 8 developed restenosis (2 had immediate operation; 4 had ineffective repeat valvotomy with subsequent operation; 2 had follow-up evaluation only), and 3 required a surgical shunt because of poor right ventricular compliance (1 died postoperatively; 1 remains cyanotic; 1 underwent right ventricular surgery for restenosis).

Pulmonary valve atresia. Early results. Balloon valvotomy was successful in 9 of the 15 patients with pulmonary valve atresia but failed in 6, in whom perforation or dilation of the pulmonary valve was impossible despite the use of radiofre-
Perforation was evacuated in two. One newborn died after infants in whom the procedure failed died 24 h later from incessant atrial flutter. The other five patients underwent balloon valvotomy and did not require further intervention, and three others required a shunt because of poor right ventricular compliance (two had their shunt embolized 1 year later because it was no longer needed).

The other four of the nine effective valvotomies were ultimate failures. In addition to the two patients who died after the surgical shunt, two others developed restenosis and, after ineffective repeat valvotomy, underwent surgical relief of the right ventricular outflow tract obstruction.

Discussion

Anatomy and pathophysiology. Many newborns with pulmonary valve atresia and an intact ventricular septum have an extremely hypoplastic right ventricle. Relief of the outflow tract obstruction is hopeless, and these patients have to be considered as having a univentricular heart. However, in a large minority, the right ventricle has three good-sized parts, including a complete outflow tract. The only difference from critical pulmonary valve stenosis is an imperforate membrane instead of a dome topped by a tiny hole. The anatomy, pathophysiology and implications for therapy are similar in both conditions. Most of these patients urgently need a pulmonary blood supply through the ductus arteriosus. In other words, infusion of prostaglandin is mandatory or strongly recommended to maintain ductal patency in these newborns as a first therapeutic step.

The second step is to relieve the right ventricular obstruction. However, the various surgical techniques that have been proposed are associated with significant mortality and morbidity (3,8). Furthermore, if a surgical shunt is required, it may distort the pulmonary artery. Therefore, percutaneous balloon valvotomy has been extended to newborn infants as an alternative to operation.

Balloon valvotomy. Feasibility. Even in a sophisticated, well equipped laboratory and with a skilled patient operator, balloon valvotomy is demanding and may be impossible to perform, as it was in 16 (16.5%) of our 97 patients. This failure rate is higher than that later in life, though during our study the failure rate tended to decrease. This decrease in failure rate may be explained not only by improved training of the operator, but also by improvement of materials (catheters and balloons) as well as new methods, such as the laser (12–14) and radiofrequency currents (15). In five patients, by using radiofrequency current, we were able to cross valves that probably could not have been passed through without this method.

Safety. Balloon valvotomy is doubtless longer and more complex in newborns than in older children. Manipulations and exchanges of catheters and guides are numerous. However, we consider that a total of three deaths directly related to the procedure in 97 attempts is a tolerable rate for a learning curve. Many nonfatal complications occurred during our study, but none had long-lasting adverse effects, although some resulted in a prolonged hospital stay, particularly for the three patients with enterocolitis.

Another safety factor that concerned us was not crossing the ductus arteriosus with the guide wire or a catheter. Although it is certainly easier to drive the dilation balloon into...
place if the guide wire crosses the ductus (16), we used this technique only rarely. We believed that ductal manipulations with stiff or sharp catheters and wires might result in unexpected closure refractory to high doses of prostaglandins because it did occur in four of our patients.

**Successful valvotomy. Effectiveness.** Unlike safety, effectiveness of valvotomy is usually not a problem in newborns with pulmonary valve stenosis or pulmonary valve atresia. In the newborns with a successful balloon valvotomy, 68 (94%) of the 72 with pulmonary valve stenosis and 9 (100%) of the 9 with pulmonary valve atresia (100%) had an effective valvotomy. An important factor for effectiveness seems to be balloon size; some investigators even recommend the use of oversized balloons (17).

**Right ventricular compliance.** The most important long-term factor affecting ultimate success is right ventricular compliance, determined partly by right ventricular size. However, in our experience, size did not always correlate with compliance. In some cases, right ventricles initially considered not very hypoplastic remained poorly compliant after relief of the obstruction, more often in pulmonary valve atresia than pulmonary valve stenosis. This poor compliance may have resulted from myocardial ischemia and fibroelastosis caused by suprasystemic right ventricular pressure. Poor compliance results in poor ventricular filling, right to left atrial shunting and cyanosis. We tried unsuccessfully to improve compliance by giving oral propranolol in a few cases. However, we believe that no medical treatment can improve right ventricular compliance. The only ways to overcome the problem are to place a surgical shunt or, as we currently do, take a course of watchful waiting while infusing prostaglandins to maintain ductal patency for as long as 3 weeks. In many cases, compliance improves, and discontinuation of prostaglandins is well tolerated. If cyanosis is still too severe after 3 weeks (hemoglobin oxygen saturation <80%), we perform a surgical shunt. Ideally, the shunt should become useless after a few months, when right ventricular compliance improves. Then, if the shunt does not clog spontaneously after discontinuation of any platelet aggregation-inhibiting drug, it can be removed either by operation or by transcatheter embolization.

**Conclusions.** Balloon valvotomy is now feasible in most newborns with critical pulmonary valve stenosis or favorable forms of pulmonary valve atresia. In the latter, the use of radiofrequency currents seems to be easier and safer than the “wire-needle technique.” In our experience, the related mortality rate is low, but it is still a demanding technique, with a 16.5% rate of nonfatal complications in our study. A highly qualified and trained operator and well-equipped laboratory are thus required to eliminate the difficulties and potential hazards of the procedure. When successfully achieved, balloon valvotomy is almost always effective in relieving the outflow obstruction, and restenosis is relatively rare. Finally, only when right ventricular compliance allows forward flow through the tricuspid valve can the patient improve and the procedure considered an ultimate success. Forward flow may occur spontaneously within several days or weeks, but, if not, a surgical shunt is required. Because any type of operation was completely avoided in 53 (74%) of our 72 patients with pulmonary valve stenosis (CI 64% to 84%) and in 2 (22%) of our 9 pulmonary valve atresia patients (CI 5% to 49%) in whom balloon valvotomy could be performed, we think that these results are encouraging. If one considers valvotomy as ultimately successful in the 5 patients who required only a temporary surgical shunt and whose pulmonary valve obstruction was relieved by the sole balloon valvotomy, then the rates of ultimate success for the patients who underwent balloon valvotomy reached 76% for pulmonary valve stenosis (55 of 72 patients, CI 66% to 86%) and 55% for pulmonary valve atresia (5 of 9 patients, CI 28% to 80%). Therefore, we conclude that balloon valvotomy should be considered a valid alternative to conventional operation for critical pulmonary valve stenosis and for favorable variants of pulmonary valve atresia in the newborn infant.

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