Percutaneous balloon valvuloplasty was attempted in 10 newborn infants with critical aortic valve stenosis and severe congestive heart failure. Three had a very small left ventricle and aortic anulus. In one infant, the aortic valve could not be passed, and in another infant, a technical error resulted in severe valvular damage, aortic insufficiency and death. Among the eight patients who had effective dilation, the stenosis was relieved in seven as assessed by a significant decrease in transvalvular pressure gradient, improvement of left ventricular contraction and eventual inversion of the ductal shunting. The procedure failed in the only patient whose dilation was performed with an undersized balloon.

Aortic insufficiency occurred in three infants and was severe (perforated cusp) in one, moderate in one whose valve was dilated with an excessively large balloon and mild and transient in one. None of the three infants with a very small left ventricle recovered (two died and one underwent cardiac transplantation). Among the seven infants with a left ventricle of acceptable size, three underwent subsequent aortic valvotomy; one of these died and two had good results. The remaining four are doing well 16 ± 5 months later (mean ± SD) with mild to moderate residual aortic stenosis and normal left ventricular function.

In conclusion, percutaneous balloon valvuloplasty is an acceptable alternative to surgery in neonates with critical aortic valve stenosis. Incidence of complications and good relief of the obstruction depend on a careful technique. Immediate results are similar to those of surgery. Late prognosis depends on the quality of the left heart structures.

(J Am Coll Cardiol 1989;13:1101-5)

Critical aortic valve stenosis in the neonate is a life-threatening malformation. Infants present with intractable congestive heart failure with severe left ventricular dysfunction and often mitral valve abnormalities and endocardial fibrosis (1-3). Medical therapy is only palliative and, until recently, infants diagnosed with critical aortic stenosis at this institution underwent urgent surgery unless the left ventricle or aortic anulus was very small. Either a closed (4,5) or open heart aortic valvotomy (6,7) was performed. This procedure is often disappointing and associated with high operative mortality and morbidity (4-8); only a few centers (9,10) report a mortality rate <20%. Given the good results of balloon catheter dilation for pulmonary valve stenosis (11) or aortic valve stenosis in children (12-19), we undertook a prospective trial of percutaneous balloon valvuloplasty in neonates with critical aortic stenosis. This report describes the results in our first 10 patients 1) to assess the effectiveness and risks of the procedure in critical aortic stenosis in newborns, and 2) to compare the results with those of surgical valvotomy in the same age group.

Study patients. Between August 1985 and December 1987, 10 consecutive neonates (2 to 34 days old, 6 < 6 days old) with critical aortic valve stenosis underwent aortic valvotomy at Hôpital Necker/Enfants-Malades. All were in severe congestive heart failure and two had required artificial ventilation support before admission. Medical treatment including an infusion of prostaglandin E₁ was initiated during the diagnostic evaluation.

Diagnosis, suspected on physical examination, chest roentgenogram and electrocardiogram (ECG), was confirmed by echocardiography and Doppler study. All patients had valvular aortic stenosis with an aortic anulus 6 to 9 mm in diameter (Table I). The left ventricle was hypokinetic in all but one patient; the mean shortening fraction was 18% (±13%) (SD) in the seven patients studied by M mode...
Correlation of left ventricular end-diastolic dimension and pressure in nine newborn infants before aortic valvuloplasty. The dimensions were expressed as minor diameters measured by M-mode echocardiography. The pressures were measured at cardiac catheterization.

![Graph showing correlation of end-diastolic minor diameter and pressure](image)

Table 1. Anatomic, Functional and Follow-Up Data in 10 Patients Undergoing Balloon Aortic Valvuloplasty

<table>
<thead>
<tr>
<th>Patient</th>
<th>Diameter (mm)</th>
<th>LV Shortening Fraction (%)</th>
<th>SP Gradient (mm Hg)</th>
<th>Surgery (days after dilation)</th>
<th>Final Outcome</th>
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</table>

*S*Small left ventricle with right to left ductal shunting. Anu = aortic anulus; Al = aortic insufficiency (— none, +tr = mild and transient, + = moderate, +++ = severe); Bal = balloon; Post = after dilation; Pre = before dilation; LV shortening fraction = shortening fraction of the left ventricle (M mode echocardiography); SP Gradient = systolic pressure gradient across the aortic valve.

Echocardiography. The size and end-diastolic pressure of the left ventricle in nine patients are shown in Figure 1.

Three of the 10 newborn infants had what we usually considered to be inoperable aortic stenosis with a very small left ventricle (end-diastolic diameter < 12 mm) and high end-diastolic pressures (18 to 21 mm Hg). They also had a patent ductus shunting from the pulmonary artery to the ascending aorta on Doppler study. The remaining seven infants had a reasonably normal left ventricular size. The mean systolic pressure gradient across the aortic valve in the 10 infants was 61 mm Hg (± 24). No patient had coarctation of the aorta or aortic insufficiency, but four had mitral valve abnormalities.

**Balloon dilation procedure.** A venous approach was used for catheterization with passage through a patent foramen ovale into the left ventricle whenever possible. In these instances, simultaneous left ventricular and aortic pressures were obtained with a 3.7F catheter inserted percutaneously and passed retrograde over a 0.028 or 0.020 in. (7.1 or 5.1 cm) Schneider steerable guide wire (Schneider Medintag) and passed retrograde over a 0.028 or 0.020 in. (7.1 or 5.1 cm) Trefoil balloon, was prepared. All guide wires were 15 mm long.

The patient was then administered an infusion of 300 mg/kg body weight of heparin sulfate for anticoagulation. If the guide wire had not been placed previously, it was passed across the aortic valve. The balloon catheter was then passed over the guide wire leaving it protruding into the left ventricle to stabilize the catheter. The balloon was inflated by hand just until the "hourglass deformity" disappeared. Once this goal was achieved the balloon was not reinflated. If more than one inflation was needed, stabilization of heart rate and blood pressure was required before repeat dilation was attempted. Duration of inflation was very short—the time it takes to inflate and deflate the balloon by hand—approximately 5 s. There were no balloon ruptures.

After valvuloplasty, the Schneider catheter was replaced over the guide by the previous 3.7F catheter and pressures were measured in the aorta and left ventricle either simultaneously, when possible, or through pullback. A left ventriculogram and aortogram were then performed in the left and right anterior oblique views. The heparinization was re-
versed with 300 mg/kg of protamine sulfate. Femoral and lower limb pulses were checked and, if they had been abolished, heparinization was started again for 24 h. If the pulses were still abolished, fibrinolytic therapy with streptokinase was applied.

Results

Technical problems and immediate results (Table 1). In Patient 1, dilation was not performed because the aortic valve could not be passed. In Patient 4, the guide wire pierced the aortic valve and dilation was performed within the cusp itself. This patient died 2 days after the procedure from rapid onset of severe aortic insufficiency. He had a small left ventricle (10 mm) and aortic anulus (6 mm) confirmed at autopsy. A hole through which a 5 mm bougie could be passed was noted in the right anterior cusp. The actual valve orifice was untouched and could not be entered by a 2 mm probe.

Severe and persistent bradycardia occurred immediately after balloon deflation in Patients 2 and 6, who required intensive care and artificial ventilation. In Patient 2, the rhythm was junctional and the patient underwent pacing for 3 h. Patient 6 had, in addition, profound bleeding at the arterial puncture site; presumably, the femoral artery was torn during catheter withdrawal. This infant received transfusions, was given additional doses of protamine sulfate, and the next day the femoral pulses were normal. Thus, both patients recovered.

A femoral pulse was not present immediately after the procedure in 5 of the 10 neonates. It returned 4 and 7 h, respectively, after heparin therapy in two infants; in another two, it reappeared a few hours after thrombolytic therapy and in the last infant, it did not reappear despite thrombolytic therapy.

Among the eight infants whose valve was diluted, the mean pressure gradient across the valve decreased in six from 61 ±24 to 20 ± 14 mm Hg (p < 0.005) after dilation (Table 1, Fig. 2). This decrease was associated with an increase in flow through the valve, as suggested by improvement in contraction of the left ventricle and changes in ductal flow. The shortening fraction of the left ventricle increased from 18 ± 13 to 37 ± 15% (p < 0.005) after dilation. In the three patients with a very small left ventricle, the right to left shunt in the ductus arteriosus was reversed to a left to right shunt. In the two remaining infants, the decrease in pressure gradient was insignificant. One (Patient 2) probably had an effective dilation because he had a very low flow and pressure gradient before the dilation and dramatically improved left ventricular function after dilation. The other (Patient 9) had apparently ineffective dilation and he was the only patient in whom dilation was performed with a balloon of inadequate size (6 mm balloon for an 8 mm aortic anulus).

Aortic insufficiency occurred in three neonates, in one (Patient 4), as discussed before, because of a perforated aortic cusp. The second patient (Patient 7), the only one treated with a balloon of excessive size, had significant regurgitation noted in the postdilation aortogram. The third patient (Patient 3) had mild and transient regurgitation, not found 1 week later on Doppler echocardiography.

Patient outcome and midterm results. None of the three neonates (Cases 1, 4, and 8) with a very small left ventricle and small aortic anulus recovered. One (Patient 4) died from the aortic cusp perforation mentioned. The second (Patient 8) died 3 weeks after the procedure in severe congestive heart failure. Autopsy confirmed the diminutive left ventricle with an abnormal mitral valve, endocardial fibroelastosis and a stenotic bicuspid valve with incomplete relief of the commissural fusion. The third patient (Case 5) remained in congestive heart failure and underwent heart transplantation 1 month after the dilation with an excellent result 18 months later. At anatomic examination, this heart revealed a hypoplastic fibrotic ventricle with mitral stenosis and severe residual aortic stenosis with a split monocusp.

Among the seven patients with a left ventricle of acceptable size, three underwent open heart aortic valvotomy. Patient 1 in whom we could not pass the aortic valve was operated on at the age of 2 days and died in the immediate postoperative period. He had a monocusp aortic stenosis with a fibrotic left ventricle. In the second patient, treated with a balloon 2 mm smaller than the aortic anulus, there was a residual gradient of 52 mm Hg. The surgeon found a bicuspid valve with no tear in the commissure or valve. The transvalvular gradient decreased to 40 mm Hg and this patient is doing well 6 months after the operation, with good left ventricular function (shortening fraction 43%) despite a residual gradient of 50 mm Hg but no aortic insufficiency.
The third patient (Case 10) was operated on 15 days after an apparently effective valvotomy (residual transvalvular gradient of 20 mm Hg) because the left ventricle was still hypokinetic with congestive heart failure. The surgeon found a bicuspid aortic valve with commissure that had been split to the aortic ring, and he did not perform any surgery on the valve. The postoperative gradient did not change, but the patient improved progressively and is doing well 14 months later with a transvalvular gradient of 55 mm Hg and a hyperkinetic left ventricle (shortening fraction 44%).

The remaining four infants were treated with balloon aortic valvotomy alone. They all survived and are doing well with no medication 6 to 22 months after the dilation (mean 16 ± 5). They have mild to moderate residual aortic stenosis with a mean aortic gradient of 39 ± 6 mm Hg as estimated by Doppler study and shortening fraction of the left ventricle of 49 ± 7%. One had moderate and well tolerated aortic regurgitation.

Discussion

This report describes an early learning experience with a number of patients that is too small and a follow-up that is too short to draw definitive conclusions. However, our results show that percutaneous balloon valvuloplasty is usually possible, safe and effective in neonates with critical aortic stenosis in heart failure. With use of the femoral artery approach, we were able to enter the left ventricle in 9 of the 10 patients. Our success rate is greater and the maneuver is quicker for critical aortic stenosis than for critical neonatal pulmonary stenosis. In addition, we have learned through this series about technical details that may improve the quality and safety of the procedure.

Complications of the procedure and their prevention. Bleeding, thrombosis, and arterial spasm are potential problems. Because we used a dilator before introducing the smallest catheter possible for adequate balloon size, we have fewer difficulties in entering the femoral artery and fewer complications. We remain committed to providing significant anticoagulation during the procedure and prompt fibrinolytic therapy after its completion if femoral artery patency seems compromised.

The aortic valve perforation that occurred in our fourth patient prompted two changes in our technique. First, when it is difficult to enter the left ventricle, we do not use the 0.028 in. guide wire to cross the aortic valve but the more supple Schneider steerable guide wire. Second, we now use two-dimensional echocardiography to visualize the guide wire within the valve orifice before dilation. If the wire cannot be positioned properly, we do not dilate but perform surgery.

After the two episodes of profound bradycardia that were presumably due to vagal stimulation secondary to the sudden increase in left ventricular pressure, we added atropine in our premedication and such episodes did not reappear. The use of a trefoil catheter that leaves some aortic flow as the balloon is inflated may have played a role in this improvement. We also consider it important to maintain the ductus arteriosus wide open with an infusion of prostaglandin E1 and to dilate only once and briefly. Furthermore, the systematic venous approach allows one to pace the patient's heart temporarily.

We believe that aortic insufficiency has been related to balloon diameter. Only one of our patients had a balloon diameter larger than the anulus, and this infant had moderate aortic regurgititation after dilation. When the balloon was well positioned and its diameter not larger than the anulus, the incidence of aortic insufficiency was low (one mild and transient case in seven patients), this low incidence may also be due to our policy of performing brief dilations that are not repeated once waisting of the balloon has disappeared. In this way we limit the risks of balloon rupture that may lead to aortic wall damage (20); there were no balloon ruptures in our series. We agree with Yeager (21) that the lowest possible pressures and shortest possible inflation-deflation cycles should be used to avoid both aortic insufficiency and the marked hemodynamic changes that occur with obstruction of the left ventricular outflow tract. With the newer trefoil balloons, continued blood flow across the valve may still be possible, thus diminishing this problem (22). Apart from these precautions, it is likely that the use of a small balloon may be ineffective, as we experienced in the only patient treated with a balloon 2 mm smaller than the diameter of the aortic anulus. We therefore recommend that the balloon diameter should not be greater than the anulus estimate but not less than 1 mm below it.

Mortality. The mortality rate in the entire series was 30%. Considering that three of the four failures (three deaths and one transplantation) were likely due to associated anomalies of the left ventricle, the balloon dilation in the other seven patients had a mortality rate of 15% (1 patient) and avoided surgery in 65% (four patients). This result compares favorably with surgical treatment. Except for the series of Messina et al. (9) in which a 9% mortality rate was reported, open heart valvotomy performed with cardiopulmonary bypass has long been used; mortality rates have been 50% to 68% in infants <8 weeks old (6-8). Valvotomy using inflow occlusion in a selected group of neonates with a good sized left ventricle has an associated mortality rate of 13% (10). Castaneda and Norwood (23) reported similar results with this technique though it is not specified whether their patients were neonates. Closed valvotomy with blunt dilation has been described (4) and we have used this technique with a mortality rate of 35% (5).

Evaluation of relief from stenosis. It is very difficult to quantify the degree of aortic stenosis in neonatal critical aortic stenosis because cardiac output is low and unstable. In these situations, the transvalvular pressure gradient is a
very poor indicator of the stenosis. The evaluation is even more difficult when the ductus arteriosus is open, as a left to right arterial shunt increases the flow through the aortic valve, whereas a right to left arterial shunt decreases it dramatically. Results are therefore subjective, based on the simultaneous changes in transvalvular gradient, size and contraction of the left ventricle, and analysis of ductal flow. An unquestionable relief in the stenosis occurs when the transvalvular pressure gradient decreases while the contraction of the ventricle increases or when the right to left ductal shunting changes to left to right shunting. With the use of these criteria, the immediate improvement occurred unequivocally in 6 of the 10 patients, possibly in 2 others, whereas there was a clear failure in 2 (1 by not passing the valve and one by dilating it with a small balloon). These results are similar to our results with surgery (5). In addition, it is interesting that in the infant who was operated on after adequate dilation but who had residual congestive heart failure, the surgeon found a split commissure that he could not open further.

**Aortic insufficiency** occurred in three patients after dilation, but it was likely related to technical errors in two of them. Our experience is too small to state whether this factor is going to be a common problem with this procedure.

**Conclusions.** Our follow-up period is too short to permit prediction of the long-term results in relation to rate of restenosis. However, these preliminary results are encouraging and lead us to proceed with percutaneous balloon valvuloplasty as the first intervention in neonates with critical aortic stenosis.

**References**