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Modified Technique for Balloon Valvuloplasty of Critical Pulmonary Stenosis in the Newborn

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Objectives. **We report our experience in eight consecutive neonates who underwent attempted balloon dilation as an initial therapy for critical valvular pulmonary stenosis, and we review in detail technical modifications that improved the success rate.**

Background. **Balloon dilation of the pulmonary valve has become the treatment of choice for valvular pulmonary stenosis in children and adults. There are few reports of its effectiveness in critical pulmonary stenosis in the newborn. In this setting, application of the technique of balloon dilation has been limited by the ability to advance the necessary guide wires and catheters across the stenotic, often near-atretic, pulmonary valve.**

Methods. **The pulmonary valve was crossed in all patients. When this could not be accomplished with an end-hole catheter, a soft guide wire was advanced directly across the pulmonary valve through the end-hole catheter positioned in the right ventricular outflow tract below the valve. Initial predilation was achieved in all patients by using a coronary dilation catheter in an effort to facilitate introduction of the definitive balloon dilation catheter. Definitive dilation with a balloon diameter of ≥110% of the diameter of the pulmonary valve annulus was possible in six patients.**

Results. **Right ventricular pressure declined from a mean value of 108 ± 32 mm Hg to a mean value of 49 ± 11 mm Hg after balloon dilation, with no change in heart rate or aortic pressure in these six patients after definitive balloon dilation.**

Conclusions. **The results of this small series suggest that critical valvular pulmonary stenosis in the newborn can be successfully treated by transluminal balloon valvuloplasty.**

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Percutaneous transluminal balloon valvuloplasty has become the procedure of choice in children with congenital valvular pulmonary stenosis (1–7). Critical valvular pulmonary stenosis with an intact ventricular septum usually presents in the newborn period in association with significant clinical cyanosis due to right to left atrial shunting and always requires urgent intervention. Until recently, therapy was exclusively surgical pulmonary valvotomy (8,9). In some patients, an aortopulmonary shunt was required to augment pulmonary blood flow when this was critically limited by right ventricular cavity hypoplasia, right ventricular noncompliance or severe tricuspid insufficiency. To date, treatment with balloon valvuloplasty of critical pulmonary stenosis in the newborn is limited by technical considerations. We report our experience in eight consecutive neonates with critical valvular pulmonary stenosis who underwent attempted balloon valvuloplasty as primary therapy, and we describe modifications in the technique that allowed an improved success rate.

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Methods

The study subjects comprised all newborns admitted from March 1989 to February 1992 to the State University of New York Health Science Center in Syracuse, New York with the diagnosis of critical valvular pulmonary stenosis with an intact ventricular septum. The diagnosis was made before birth in two infants. There were eight infants, seven male and one female. They were 1 to 5 days old (weight 3.2 to 4.3 kg) at the time of cardiac catheterization. All had clinical cyanosis, with a mean room air arterial partial pressure of oxygen Po_2 of 33 ± 5 mm Hg. After the diagnosis was confirmed by two-dimensional echocardiographic and color Doppler evaluation, prostaglandin E_1 infusion was initiated in all. Findings at presentation are summarized in Table 1.

All infants underwent cardiac catheterization with attempted balloon valvuloplasty after their condition had stabilized and parental consent had been obtained. Before catheterization, two infants underwent tracheal intubation to stabilize ventilation. The femoral vein was entered percutaneously in all patients, and a venous sheath (USCI) was introduced. In six infants, an umbilical artery catheter was used to monitor aortic pressure, and in two patients a femoral artery catheter (4F UMI Teflon pigtail) was inserted for this purpose. Right and transatrial left heart hemodynamic measurements and saturations were obtained, and right and left ventricular angiography was performed in all.

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Table 1. Pertinent Patient Data

Pt No.	Age (days)/ Gender	Wt (kg)	Pao_2 (mm Hg)	PV Annulus Diameter		Balloon Diameter (mm)	RV Area (cm^2)	TV Annulus Diameter		TR	Hospital Stay (days)
				Echo (mm)	Cath (mm)			Echo (mm)	Cath (mm)		
1*	4/M	4.5	33	7	5	4	2.2	9	12	4+	24
2	5/F	3.2	34	7	7	8	1.9	9	9	—	7
3	3/M	4.3	33	9	7	10	4.7	10	10	—	5
4*	1/M	4.3	—	9	6	5	6.8	10	13	1+	18
5	2/M	3.3	27	8	7	10	2.3	11	11	2+	9
6†	1/M	3.9	25	5	7	10	1.5	11	12	4+	18
7	1/M	3.4	39	7	7	8	3.3	11	11	4+	9
8	1/M	4.2	38	9	8	10	6.3	15	12	4+	5
							3.6 ± 2.1				

*Required surgical valvotomy after predilation with a coronary dilation catheter. †Required systemic to pulmonary artery shunt after successful balloon valvuloplasty with an 8- and, subsequently, a 10-mm diameter balloon. Cath = catheterization; Echo = echocardiography; Pao_2 = room air arterial partial pressure of oxygen; Pt = patient; PV = pulmonary valve; RV = right ventricular; TR = tricuspid regurgitation; TV = tricuspid valve; Wt = weight.

The size of the pulmonary valve annulus was assessed from the lateral angiogram and was then compared with the echocardiographic measurement obtained before catheterization to guide in appropriate balloon size selection. Balloon dilation catheter diameters were chosen to be 110% to 130% of the measured pulmonary valve annulus diameter.

The pulmonary valve was crossed in all infants with either a 4F or a 5F balloon end-hole catheter (Arrow). In four infants, a soft J-tip 0.016-in. (0.041 cm) steerable (USCI) guide wire was used to cross the valve after the end-hole catheter had been positioned in the infundibular region of the right ventricle (catheter tip directed somewhat rightward and posterior), as one would for normal catheter passage across the pulmonary valve. With the wire in place, the catheter was gently coaxed across the pulmonary valve.

The end-hole catheter was removed, leaving the 0.016-in. guide wire positioned in either a branch pulmonary artery (five of eight infants) or the descending aorta through the ductus arteriosus (three of eight infants). A low profile coronary dilation catheter (4-mm diameter in three infants, 5-mm diameter in five; length either 2.0 or 2.5 cm) was advanced over the guide wire, and predilation with two or three inflations was performed. The coronary balloon dilation catheter was withdrawn, leaving the guide wire in position. The venous sheath was changed as necessary to allow introduction of a larger end-hole catheter (7F balloon end-hole catheter [Arrow] in six patients, 4.1F multipurpose [Cordis] catheter in two patients), followed by a 0.035-in. (0.089 cm) J-tip Teflon-coated exchange guide wire (Cook) positioned in the branch pulmonary artery in the initial four patients and in the descending aorta by way of the ductus arteriosus in the last four patients.

The end-hole catheter was removed, leaving the 0.035-in. guide wire in place, and the preselected balloon catheter was introduced. This could not be accomplished in two infants early in our experience. In six infants the 0.035-in. guide wire was successfully maintained in position, and the balloon dilation catheter was positioned across the pulmonary valve. Passage of the catheter was greatly facilitated in the last four infants with the guide wire anchored in the descending aorta through the ductus arteriosus.

Use of a low profile dilation catheter (NuMed) allowed the venous sheath to remain in place in the last three infants. Valvuloplasty was carried out with elimination of the balloon waist for two or three inflations with dilute contrast agent lasting ≤10 s each (8-mm diameter in three patients, 10-mm diameter in four patients, all with a 3.0-cm–length balloon).

Statistical analysis. Data are presented as mean value ± 1 SD. The paired Student *t* test was used to compare predilation and postdilation cardiac hemodynamic data.

Results

Definitive balloon valvuloplasty was accomplished in six of the eight infants, with a mean ratio of balloon diameter to pulmonary valve annulus diameter of 1.3 ± 0.2 (range 1.1 to 1.4). Total fluoroscopy time averaged 60 ± 27 min in these six infants. Hemodynamic findings are summarized in Table 2. Right ventricular systolic pressure decreased from a mean of 108 ± 32 mm Hg before balloon valvuloplasty to a mean of 49 ± 11 mm Hg ($p < 0.001$) immediately after valvuloplasty. The ratio of right ventricular systolic pressure to aortic pressure also decreased from a mean of 1.44 ± 0.25 to 0.66 ± 0.12 ($p < 0.001$). No significant difference was noted when comparing heart rate or aortic systolic pressure before and after valvuloplasty.

Pulmonary valve annulus diameter determined by echocardiography was 7.5 ± 1.5 mm and was comparable to the 7.2 ± 0.5 mm determined by angiography. In these six infants, tricuspid regurgitation was severe in three and mild in one (as assessed by both angiography and Doppler echocardiography before catheterization). One infant (Patient 6) with severe tricuspid insufficiency required an aortopulmonary shunt for persistent profound hypoxemia due to right to

Table 2. Hemodynamic Results*

Pt No.	Before Valvuloplasty				After Valvuloplasty			
	HR (beats/min)	RVp (mm Hg)	Aop (mm Hg)	RVp/Aop	HR (beats/min)	RVp (mm Hg)	Aop (mm Hg)	RVp/Aop
1†	150	80/8	85/52	0.94	154	69/8	86/40	0.80
2	140	160/8	93/58	1.72	110	67/7	103/63	0.65
3	121	122/6	84/53	1.45	144	38/3	77/53	0.49
4†	140	102/20	82/46	1.24	140	73/20	82/50	0.89
5	155	105/17	75/42	1.40	150	42/7	65/42	0.65
6‡	150	85/13	60/32	1.42	150	48/18	63/35	0.76
7	156	107/13	66/37	1.62	175	58/19	70/40	0.83
8	150	66/5	66/38	1.00	150	40/7	60/40	0.67
8/8	145 ± 11	103 ± 29	76 ± 12	1.35 ± 0.27	147 ± 18	54 ± 14	76 ± 14	0.72 ± 0.13
6/8	145 ± 13	108 ± 32	74 ± 13	1.44 ± 0.25	147 ± 21	49 ± 11	73 ± 16	0.68 ± 0.12
					NS	$p < 0.01$	NS	$p < 0.001$

*Statistical analysis was performed comparing heart rate, right ventricular pressure, aortic pressure and ratio of right ventricular to aortic pressure before and after valvuloplasty in the six patients who had definitive balloon valvuloplasty. Values presented are mean value ± 1 SD. †Required surgical valvotomy after predilation with coronary dilation catheter. ‡Required systemic to pulmonary artery shunt after successful balloon valvuloplasty with an 8- and, subsequently, 10-mm diameter balloon. Aop = aortic pressure; HR = heart rate; Pt = patient; RVp = right ventricular pressure; RVp/Aop = ratio of right ventricular to aortic pressure.

left atrial shunting after successful balloon dilation of the valve.

Duration of hospital stay appeared to be directly related to the need for surgical intervention. In five of the six infants with successful balloon valvuloplasty, the duration of the hospital stay ranged from 5 to 9 days. The infant who required an aortopulmonary shunt for severe persistent cyanosis had a hospital stay of 18 days. In two infants without definitive balloon valvuloplasty, the hospital stay was 24 and 18 days, respectively.

Definitive balloon valvuloplasty could not be performed in two infants. Predilation was performed in both with a 4-mm diameter coronary balloon dilation catheter. In Patient 1, the 0.035-in. guide wire was not stable enough in the branch pulmonary artery to allow removal of the 7F end-hole catheter, and the wire fell back into the right ventricle. Further attempts to cross the pulmonary valve were unsuccessful in this patient, who subsequently underwent successful surgical pulmonary valvotomy. In Patient 4, the right ventricular outflow tract was perforated with the guide wire after the wire fell back into the right ventricle from a branch pulmonary artery during attempts to advance the definitive balloon dilation catheter. Minimal hemopericardium was evacuated in the operating room without complication at the time of open pulmonary valvectomy. The postoperative course in both Patients 1 and 4 was complicated by severe hypoxemia due to massive right to left atrial shunting that gradually resolved.

Follow-up. Doppler echocardiographic evaluation was available at a mean of 13 months (range 2.5 to 34) after valvuloplasty in all patients. The mean estimated right ventricular outflow gradient was 16 ± 15 mm Hg (range 0 to 40), with mild pulmonary regurgitation present in all six patients with successful balloon valvuloplasty. No significant tricuspid regurgitation was present in these six patients. In Patient 6, evaluation 6 months after intervention demonstrated no residual pulmonary stenosis, a patent aortopulmonary shunt and minimal right to left atrial shunting through a tiny atrial patency. Cardiac catheterization in Patient 6 at 12 months of age revealed no residual pulmonary stenosis and a patent right modified Blalock-Taussig shunt that was subsequently coil embolized when the patient was 17 months old.

Of the two patients who had surgical repair, Patient 1 underwent repeat balloon valvuloplasty and closure of an atrial septal defect with an umbrella device 4 months after the initial procedure. At 25 months after the initial repair, the estimated right ventricular outflow tract gradient was 32 mm Hg, with moderate pulmonary regurgitation. Patient 4 had no residual right ventricular outflow tract gradient, with moderate pulmonary regurgitation noted 15 months after the initial surgical repair. No clinical evidence of venous stasis or diminished lower extremity pulse was noted at follow-up evaluation in any patient.

Discussion

In newborns with critical valvular pulmonary stenosis present with profound cyanosis, urgent treatment is necessary to establish pulmonary blood flow. Initial stabilization is achieved with prostaglandin E_1 infusion after Doppler echocardiographic diagnosis (10–12). Until recently, treatment has usually been surgical, with significant morbidity and mortality (13).

Up to this time, there have been only sporadic reports of balloon dilation of the pulmonary valve in the newborn because of the rarity of the lesion and the technical difficulty of the procedure (14,15). In our series, successful balloon valvuloplasty resulted in an immediate decrease in right ventricular systolic pressure as well as clinical improvement,

with the need for only a relatively short hospital stay in these critically ill infants.

Transluminal balloon valvuloplasty appears to be an effective alternative treatment for critical pulmonary stenosis with an intact ventricular septum in the newborn period. However, the procedure is technically demanding, and previous experience suggests a moderate failure rate (14,15). Technical problems in two of the initial four patients led to a modification of the technique so that valvuloplasty was readily accomplished in the last four patients.

Technical modifications that led to an improved success rate include 1) use of a soft-tipped guide wire to cross the pulmonary valve directly, with the catheter positioned in the right ventricular outflow tract; 2) stabilization of the guide wires across the pulmonary valve in the descending aorta through the ductus arteriosus to facilitate passage of subsequent catheters and wires; and 3) predilation of the valve with low profile coronary balloon dilation catheters to facilitate passage of the catheters and guide wires necessary for definitive dilation. The availability of lower profile dilation catheters (NuMed) capable of being introduced through a venous sheath also greatly facilitated the procedure.

Conclusions. The results of this small series suggest that balloon valvuloplasty can be successfully used as a primary therapy for critical pulmonary stenosis in the newborn. With careful attention to technical details, a high success rate with low morbidity is possible. Consideration should be given to selection of balloon valvuloplasty as the initial procedure in the treatment of neonates with critical pulmonary valve stenosis. However, critical evaluation of this procedure should continue to ensure that these results are indeed valid.

Addendum

Recently, a 13-day old, 4-kg white female infant had balloon valvuloplasty performed in the previously described fashion with the guide wire anchored in the descending aorta by way of the ductus arteriosus for severe pulmonary valvular stenosis. Before valvuloplasty, the right ventricular pressure was 100/5 mm Hg, aortic pressure was 78/49 mm Hg and the ratio of right ventricular to aortic pressure was 1.28. After valvuloplasty, the right ventricular pressure was 60/5 mm Hg, aortic pressure was 99/57 mm Hg and the ratio of right ventricular to aortic pressure was 0.61.

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