Short and Long-Term Outcomes after Balloon Pulmonary Valvuloplasty in Congenital Pulmonary Stenosis

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Abstract

Background- Balloon pulmonary valvuloplasty (BPV) has emerged as the treatment of choice for patients with valvular pulmonary stenosis (PS). We report here our short and long–term outcomes of BPV in 64 patients with isolated native PS.

Methods- From February 1996 to February 2006, sixty–four patients with PS (pressure gradients ≥40 mm Hg) were enrolled in this retrospective study.

Results- The hemodynamic data at catheterization revealed that the RV - PA pressure gradient before BPV ranged from 40 to 240 mmHg (mean ± SD = 93.2 ± 43.4 mmHg). The above gradient immediately after BPV ranged from 5 to 163 mmHg (mean ± SD = 30.3 ± 27.7 mmHg), and the difference was significant (p=0.0037). Twenty-three patients had regular follow–up. The duration of follow– up ranged from 1-120 months with a mean of 38.5 ± 31.3 months. The transvalvar pressure gradient during the above period ranged from 10 to 140 mmHg with a mean of 35.9 ± 27.9 mmHg and showed a significant difference (p = 0.0032) with the pressure gradients before BPV.

Conclusion- BPV provides short and mid-term relief of pulmonary valve obstruction in the majority of patients ***(Iranian Heart Journal 2010; 11 (2):25-29).***

Key words: pulmonary stenosis ■ balloon angioplasty ■ outcome

Pulmonary stenosis (PS) is rather common, accounting for 7.5 - 9% of the cases of congenital heart disease. The pathologic features of the stenotic pulmonary valve vary; the most commonly observed pathology is described as dome – shaped pulmonary valve. In the past, surgical valvotomy was the treatment of choice. Percutaneous balloon pulmonary valvuloplasty (BPV) is now the treatment of choice for congenital PS.[1]

The indications for BPV are similar to those used for surgical pulmonary valvotomy, i.e. a moderate degree of PS with a peak – to peak gradient ≥ 50 mmHg with normal cardiac index.[2] Some workers use lesser gradients (40 mmHg) or right ventricular pressure of 50 mmHg for intervention.

Immediate reduction of peak–to-peak gradient across the pulmonary valve and of RV peak systolic pressure occurs following balloon dilatation. The width of the jet of the contrast material passing through the pulmonary valve increases and surgical intervention is avoided in most patients.[1]

Most patients are discharged home within 24hr of the procedure. At mid–term follow-up (usually defined as <2 years), both catheterization measured peak–to-peak and Doppler measured peak gradient remain improved for the patients as a whole. However, restenosis, defined as gradient ≥ 50 mmHg is observed in 8-10% of patients.[3]

Patients with restenosis have been

Received Apr. 2, 2009; Accepted for publication Jun. 23, 2010
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successfully treated by performing re-dilatation with balloons larger than those used at the time of initial BPV. Two risk factors for recurrence have been identified: balloon / pulmonary valve annulus ratio <1.2 and immediate post-valvuloplasty gradient ≥ 30 mmHg.[1] At long– term follow–up (usually defined as >2 years) pulmonary valve insufficiency (PI) is noted in 40-90% of patients. The prevalence and the degree of PI increases with time; however, no one has required surgical intervention in the majority of reported studies.[4, 5]

The purpose of the present study was to report the results of short and long – term follow - up of BPV in patients with PS at a pediatric cardiology ward of a university hospital in Tehran, Iran.

Methods

The present study is a retrospective study. The medical records of all patients with discharge diagnosis of PS were evaluated. These cases had been admitted during a period of 10 years (1996-2006) to the Pediatric Cardiology Ward of Shaheed Modarres Hospital. Cases with dysplastic valves, syndromic PS, and associated CHD were excluded from the study. Patients with isolated PS and transvalvar pressure gradients ≥ 40 mmHg were entered in the study. The demographic characteristics of the patients and their clinical and paraclinical features (chest radiography, electrocardiography (ECG), echocardiography (echo), catheterization, and angiographic data, as well as follow–up profile) all were collected from their charts and depicted in a data sheet. Diagnostic catheterization and BPV were performing during the same procedure. Informed consent was obtained from the parents. The patients were sedated with a sedative cocktail, given intramuscularly half an hour before the procedure. Percutaneous femoral venous route was the entry site for all the patients. Measurement of the right ventricle (RV) and PA pressure along with peak– to - peak gradient across the pulmonary valve was performed. This peak – to - peak gradient was used to assess the severity of pulmonary valve obstruction. The selected balloon angioplasty catheter was advanced over the guide wire and positioned across the pulmonary valve. The size of the balloon was generally 1.2 to 1.3 times the pulmonary valve annulus. The balloon was inflated with diluted contrast material, while monitoring the pressure of the inflation. The inflation pressure was gradually increased until the disappearance of the balloon waist. If the result was not satisfactory, a repeat dilatation with a larger balloon (2 mm larger than the first one) was undertaken. The type of the catheter in the first half of the study period was the Bult catheter and in the second half was the Tay-Shak catheter and Zed - Med catheter. The patients were observed in an intermediate care setting overnight. Echocardiography was performed on the morning following the procedure. Clinical, ECG, and echo-Doppler evaluations were performed at 1, 6, and 12 months after the procedure and yearly thereafter. Restenosis, defined as gradient ≥40 mmHg, was evaluated during the follow-up period. Pulmonary regurgitation was graded subjectively as trivial, mild, moderate, or severe based on clinical, and / or pulsed and color Doppler echocardiographic assessments.

Data analysis was conducted using SPSS software. The continuous variables were presented as means ± standard deviation (SD). Student's *t*-test for paired data was used to compare the pressure gradients. A P value of <0.05 was considered statistically significant.

Results

During a 10–year period from February 1996 to February 2006, a total of 64 consecutive patients (35 male and 29 female) with isolated PS underwent BPV at a pediatric cardiology ward in Tehran, Iran. Chart recordings of

clinical manifestations, plain chest films, electrocardiography, echocardiography, hemodynamic data of cardiac catheterization, and follow – up profile were all available for review in these patients. The patients' age ranged from 1 to 32 years with a mean of 10.5 ± 7.7 years. Distribution of the patients' age group is as follows: 1 – 5 yr old, 18 (28.1%) cases; 6 – 10 yr old, 20 (31.2%) cases; 11 – 15 yr old, 12 (18.8%) cases; 16 – 20 yr old, 5 (7.8%) cases; and > 20 yr old, 9 (14.1%) cases. Thus 38 (59.3%) cases were young children (≤ 10 yr old). All the patients had a typical systolic ejection murmur over the left upper sternal border. Fifty-five of the 64 (85.9%) patients had right-axis deviation, and 56 (87.7%) patients showed right ventricular hypertrophy in ECG. Post-stenotic pulmonary artery dilatation (prominent pulmonary segment) was present in 18 (28.2%) patients. Echo – Doppler was performed for all the patients before catheterization. The right ventricle–pulmonary artery (RV–PA) pressure gradients by Echo Doppler ranged from 42 to 235 mmHg with a mean of 92.1 ± 45.2 mmHg. Table I summarizes the RV-PA pressure gradients before BPV at the catheterization laboratory. The RV-PA pressure gradient before BPV ranged from 40 to 240 mmHg with a mean of 93.2 ± 43.4 mmHg.

Table I. Distribution of pressure gradients before BPV at the catheterization laboratory

Pressure gradient before BPV (mmHg)	Number	Percent
40 - 80	31	48.5
81 - 120	20	31.2
121 - 160	8	12.5
161 - 200	4	6.2
> 200	1	1.6

Table II shows the distribution of the pressure gradients immediately after BPV. The RV-PA pressure gradient immediately after dilatation with balloon ranged from 5 to 163 mmHg with a mean of 30.3 ± 27.7 mmHg. There was a significant difference between the pressure gradients (PG) before and after dilatation (p = 0.0037) at the catheterization laboratory. All the patients underwent Doppler echocardiography the following morning after catheterization. The RV-PA pressure gradients by echo ranged from 16 to 158 mmHg, with a mean of 44.7 ± 25 mmHg. There was not a significant difference between the PGs immediately after BPV by angiography and PGs 24 hours after BPV by echocardiography (p = NS). Forty–one (64 %) patients were lost to follow-up and 23 had regular follow-ups. The duration of follow-up ranged from 1 to 120 months, with a mean of 38.5 ± 31.3 months. The RV-PA pressure gradients during the follow–up period ranged from 10 to 140 mmHg, with a mean of 35.9 ± 27.9 mmHg. There was a significant difference (p = 0.0032) between the PGs before BPV and PGs after a mean follow –up period of 38.5 ± 31.3 months.

Table II. Distribution of pressure gradients immediately after BPV at the catheterization lab

Pressure gradient after BPV (mm Hg)	Number	Percent
≤ 20	25	39
21 - 40	27	42.2
41 - 60	6	9.4
61 - 80	3	4.7
> 80	3	4.7

Our data indicated that 12 patients had PGs > 120 mmHg before BPV and most of them had PGs > 40 mm Hg immediately after BPV, but half of these patients (6 of 12) had gradual resolution of obstruction during the follow-up period. Four of these 12 cases were lost to follow up, one case required re-dilatation by BPV, and one case underwent surgical intervention. Nine of our cases were older than 20 years, and 5 out of these 9 presented with PGs ≥ 80 mmHg. These patients showed moderate tricuspid insufficiency and right

ventricular cardiomyopathy. All of them showed improvement with gradual decline in PG during the follow-up period. Mild pulmonary insufficiency by clinical and / or echocardiographic examination was detected in 48.2% of our patients. There was one technical error in our series. The balloon had been dilated at an improper level of the pulmonary valve in a 23–year-old boy. The problem was safely solved by repeating the procedure. Transient bradycardia and premature beats during balloon inflation was noted in many of our patients. These abnormalities returned rapidly to normal following balloon deflation. There was no mortality in our series.

Discussion

The rationale for BPV intervention has included the prevention and relief of symptoms, the prevention of secondary changes in the RV and PA, and the prevention of progression to more severe degrees of obstruction. Therefore, the indications for intervention based on natural history data might include any patients with moderate or severe obstruction, any patients with significant symptoms, and children with mild degrees of obstruction who have shown any evidence of progression at follow-up evaluations.[6] So it is not unreasonable that patients with gradients of less than 50 mmHg (mild stenosis) might be treated via BPV. Our findings showed that there were 7 cases with PGs between 40 to 50 mmHg who needed intervention using BPV due to the increasing intensity of signs, symptoms, and PGs. In our series, 33 (51.5%) patients had signs and symptoms of moderate to severe PS with PG > 80 mmHg. We demonstrated a significant reduction in the pre-dilatation gradient in 52 (81.3%) patients. Later reductions in PG occurred in 6 more patients during the follow-up. Thus, we achieved a success rate of 90.6% (58 of 64) in our cases. Early success rates for individual series ranged from 37 % to 100%, with 80 % of patients having had successful procedures.[6] In a study by Hatem et al., 189 patients with PS underwent BPV during a period of 12 years (1984 – 1996). The mean age of patients was 7.9 ± 9.2 years. The follow-up ranged from 4 to 12 years. Immediate success was observed in 148 of the 189 (78.7%) patients. Later reduction occurred in 24 more patients. So BPV was effective in 172 (91%) cases.[7] In another research by Rodriguez et al., 150 cases with PS and a mean age of 10.5 ± 11.3 years were followed for 48 ± 44 months. Immediate success was observed in 111 (74%) patients. At the end of the follow-up period, successful outcomes were achieved in 89.6%. The predictors for failure were age < 1.5 ± 1.3 years, dysplastic valve, high initial systolic gradient, and high initial systolic RV pressure.[8] Data from the large VACA (Valvuloplasty and Angioplasty of Congenital Anomalies) registry study suggest that the majority of these significant residual gradients are at the infundibular level and that the higher the degree of total obstruction before BPV, the higher the infundibular gradient immediately after BPV.[9] Infundibular obstructions regress to a great degree at follow-up, just as has been demonstrated for infundibular reaction following surgical valvotomy.[10] We administered beta-blockers for the resolution of subvalvular hypertrophy successfully.

Pulmonary valve insufficiency (PI) is noted in some patients after BPV. The frequency and severity of PI increases with time; 41-88% may develop PI at long-term follow-up. Residual PI does not require surgical intervention in most patients.[11] Data from a study in Turkey confirmed that balloon dilatation in isolated valvular PS is safe and effective and suggested that stenosis does not recur.[12] Thirty–one (48.2%) cases of our group showed trivial to minimal degrees of PI during the follow–up period. They were all asymptomatic and tolerated PI without any intervention. We did not find restenosis in our

study group. Complications during and immediately after balloon valvuloplasty have been remarkably minimal. The VACA registry reported a 0.24% death rate and 0.35% major complications rate from the 822 BPV procedures at 26 institutions.[9] Our experience showed one technical problem during the procedure, and no mortality throughout the study.

Following BPV in childhood, gradient reduction persists and RV structure grows appropriately in the majority of children. Late PI occurs commonly, but it is well tolerated. While life–long follow-up is essential, excellent outcome can be anticipated.[13] our findings showed that 41 (64%) patients were lost to follow-up; many of these patients became symptom free and so did not follow the regular schedule for out patient visits. We should emphasize our encouragement for patients' follow-up.

In conclusion, BPV is an effective, safe, and the first choice of treatment for congenital pulmonary valve stenosis. This intervention has excellent short and long-term results. Emphasis for regular follow-up and multicenter studies are suggested.

Conflict of Interest

No conflicts of interest have been claimed by the authors.

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