Mid –Term Results of Balloon Pulmonary Valvuloplasty in Children at Queen Alia Heart Institute

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ABSTRACT

Objective: To evaluate the mid-term results of balloon pulmonary valvuloplasty in children.

Methods: One hundred and thirty-two patients with a median age of three years (range 3 days to 16 years), underwent balloon pulmonary valvuloplasty between January 2000 and January 2007 at the Pediatric Cardiology Department of Queen Alia Heart Institute, Amman-Jordan. We evaluated our patients with regard to clinical symptoms, peak to peak pressure gradient across the right ventricular outflow tract before and after balloon pulmonary valvuloplasty. Thereafter, we followed them at our outpatient clinic regarding clinical symptoms, the peak instantaneous pressure gradient, the presence of pulmonary valve incompetence, right ventricular function based on echocardiographic findings, and the need for repeat balloon pulmonary valvuloplasty.

Results: There was significant reduction in the peak to peak gradient from 93.5 ± 32.3 mmHg to 26.1 ± 9.6 mmHg (P<0.001). Repeat balloon pulmonary valvuloplasty was performed in 12% of cases. Surgical intervention performed for dysplastic valve and fixed subpulmonic obstruction was done in 11.36% of cases. Echo-Doppler data showed that residual peak instantaneous gradient dropped with time from 24.4±3 to 19 ± 6 mmHg (P<0.001) on follow up range (5 months to 5.2 years). Mild to moderate pulmonary valve regurgitation was noticed in 34 patients (27.8%) at one year and in 43 patients (38%) at midterm follow up, but none had right ventricular dilatation or paradoxical interventricular septal motion.

Conclusions: The results of midterm follow up after balloon dilatation of the pulmonary valve is encouraging. Restenosis occurred in 22% of cases. Children under two years had statistically significant higher pressure gradient and more pulmonary regurgitation. The degree of pulmonary regurgitation increased with time. Longer term follow up studies should be undertaken to evaluate the significance of residual pulmonary regurgitation.

Key words: Balloon dilatation, Pulmonary stenosis, Pulmonary regurgitation

Introduction

Pulmonary stenosis accounts for about 10-12% of all cases of congenital heart diseases.1 Since 1982, after the first description of balloon dilatation of pulmonary valve by Kan et al.2-5 balloon pulmonary valvuloplasty (BPV) has become the treatment of choice for moderate to severe pulmonary valve stenosis (PS) with intact ventricular septum in all ages, and completely replaced surgical valvotomy. Several studies reported excellent acute and intermediate results,6-15 but still the long term results are to be clarified.

We conducted our study to show our intermediate results and experience in balloon pulmonary valvuloplasty in children at Queen Alia Heart Institute (QAHI) in Jordan, although the first BPV performed successfully in this institute was in November 1988.
Fig. 1. Distribution of cases according to age

Methods

From January 2000 to January 2007 at QAHI, 132 patients with a median age of three years (range 1 day-16 years), and a median weight of 11 kg (range 3.5-51Kg) underwent balloon pulmonary valvuloplasty. We classified our cases according to age below two years, two to ten years and those above ten years of age (Fig. 1). Fifty four cases were males (41%) whereas 78 (59%) were females. They presented with chief complaints of shortness of breath or exertional dyspnea, underwent balloon dilatation of the pulmonary valve (BPV). Data were collected retrospectively by reviewing their medical records, noninvasive studies and cardiac angiograms to obtain acute and intermediate results. Inclusion criteria were those patients with maximum instantaneous pressure gradient (PG) across the pulmonary valve by Echo-Doppler was ≥ 50mmHg. Dysplastic pulmonary valve which was defined as the presence of thick, immobile valve leaflets with the absence of poststenotic pulmonary valve artery dilatation was noticed in 18 cases (13.6%). Excluded cases were those with associated cardiac anomalies except those with small hemodynamically insignificant secundum atrial septal defect which was observed in 29 patients (22%) who were included. Two cases had Noonan syndrome. Informed consent was obtained from the parents.
Table I. Grading of pulmonary regurgitation by echo Doppler studies

<table>
<thead>
<tr>
<th>Grade</th>
<th>Description</th>
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<tbody>
<tr>
<td>None</td>
<td>No pulmonary regurgitation on Doppler study</td>
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<tr>
<td>Grade I</td>
<td>Pulmonary regurgitation jet width ≤ 10% of pulmonary valve annulus diameter in precordial short axis view</td>
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<td></td>
<td>No right ventricle volume overload</td>
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<tr>
<td>Grade II</td>
<td>Pulmonary regurgitation jet width 1-25% of pulmonary valve annulus diameter</td>
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<tr>
<td></td>
<td>No right ventricle volume overload</td>
</tr>
<tr>
<td>Grade III</td>
<td>Pulmonary regurgitation jet width 26-50% of pulmonary valve annulus diameter</td>
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<tr>
<td></td>
<td>No right ventricle volume overload but with or without flat septal motion</td>
</tr>
<tr>
<td>Grade IV</td>
<td>Pulmonary regurgitation jet width &gt; 50% of pulmonary valve annulus diameter</td>
</tr>
<tr>
<td></td>
<td>Right ventricle volume overload present</td>
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</tbody>
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Jet width at the origin of regurgitation jet rather than jet length was used for grading because the jet width is not influenced by pulmonary artery pressures.

Right ventricle volume overload is defined as enlarged right ventricle (> 95 percentile) and flat to paradoxical septal motion.

of each patient after fully describing the technique and the aim of the procedure. After initial hemodynamic assessment right ventricular angiography was performed, and maximum internal diameter of the pulmonary valve from hinge point to hinge point during systole was measured from the lateral projection cineangiogram and corrected for magnification. Our technique of balloon dilatation of the pulmonary valve was similar to that described in details by others.4,16-19 (Fig. 2-4). The balloon size used was 1.2 to 1.4 times the size of the measured pulmonary valve annulus on the lateral projection. We define the success rate if peak to peak pressure gradient (PG) by pull back pressure tracing post BPV was ≤ 35mmHg immediately after the procedure. All patients were given heparin in a dose of 100 Units/Kg during the procedure. The patient usually stayed in hospital for one day after the procedure, received intravenous antibiotics and was discharged after performing 2D-Echo Doppler evaluation.

Follow up evaluation as outpatient included both clinical and 2D-Echo and Doppler examination at one, three and six months and then yearly thereafter. The clinical evaluation focused on clinical signs of stenosis. The 2D-Echo-Doppler evaluation assessed the maximum instantaneous pressure gradient across the pulmonary valve, the presence of pulmonary regurgitation (PR) if it was grade I, II, III or IV according to the color jet width of regurgitant flow across the pulmonary valve as per RAO et al.’s40 (Table I) classification, which was performed in the precordial short axis view and the right ventricular function as well and finally the interventricular septal movement by the M-mode.

Statistical analysis:
All data were expressed as mean ± SD or median with range. Paired t-test was used to compare the mean right ventricular pressure and pressure gradient across the pulmonary valve before and after balloon valvuloplasty. A P value less than 0.05 was considered significant.

Results
Immediately after the initial balloon pulmonary valvuloplasty there was a significant reduction in the peak to peak gradient from 93.5±32.3mmHg to 26.1±9.6mmHg (P value <0.001) (Fig. 5). There was also a significant reduction in the right ventricular pressure (RVP) from 117.6±33.1mmHg to 50.7±9.9mmHg with a P value <0.001 (Fig. 6). Forty-five patients (34%) had re-stenosis and their immediate peak to peak maximum PG post BPV was ≥ 35mmHg, making the early success rate of 66%. Sixteen of them (12%) underwent repeat dilatation of the pulmonary valve, in a mean period of 12±4months after the initial dilatation with significant reduction of their peak to peak PG from 65±3.65 mmHg to 21±2.8mmHg (p<0.001) (Fig. 7) and their RVP dropped from 87.5±5.9mmHg to 41±3.8mmHg (p<0.001) (Fig. 8) making the success rate rise to 78%. BPV was performed for 18 cases with dysplastic pulmonary valve and was successful in seven cases making their success rate of 38.9%. Eighteen patients (13.6%) with mild to moderate infundibular stenosis of 33±18mmHg, four of them had subpulmonic obstruction before BPV and persisted after balloon dilatation, whereas the other ten showed improvement with the use of β- Blockers (propranolol of a dose of 1mg/kg/dose twice or thrice daily) which was given for two to three months. Fifteen patients (11.36%) needed surgery after BPV, eleven due to dysplastic valve and four due to fixed subvalvular obstruction. Patients below two years of age had their peak to peak graph PG pre BPV significantly higher than those who were above two years (98±35.6 mmHg vs. 86±26.5 mmHg P=0.03) (Fig. 9) and has a higher rate of re-stenosis post BPV (mean PG=75±2.2 mmHg for 27 patients below two years vs. 55±1.4mmHg for 18 patients above two years; P<0.001) (Fig. 10). Eight patients
missed their follow up. During follow up, for those who came back 126 cases, the residual peak instantaneous gradient dropped further from 24.4±3 to 19±6mmHg with P<0.001 (Fig 11).

Grade I-II pulmonary valve regurgitation was noticed in 34 (27.8%) patients at one year and in 43 (38%) patients, at mid term follow up range (range 5 months to 5.2 years), 28 patients out of 43 had grade II regurgitation, their mean PG was 75±2.2 before BPV, they had statistically significant more regurgitation (grade II) than the 15 patients who had grade I regurgitation with their mean PG being 55±1.4 and P<0.001 (Fig. 12), but neither right ventricle dilatation or impairment nor paradoxical interventricular septum motion occurred. Mortality rate of those who came back for follow up 124 (93.9%) cases was 0%, immediately and on mid-term follow up.

Discussion
Pulmonary valve stenosis is one of the common congenital heart diseases. The traditional method of treatment was surgical valvotomy until 1982, when Kan et al. introduced the technique of percutaneous balloon valvuloplasty. Since that time, it replaced the surgical option except for few exceptions: pulmonary valve hypoplasia or the presence of concomitant intra cardiac defects which need to be addressed at the same time. Although the majority of our patients with pulmonary valve stenosis were asymptomatic, our rationale for taking our patients for BPV when their peak to peak maximum PG≥50mmHg in order to prevent and relieve the symptoms, to prevent the secondary changes in the right ventricle and the pulmonary artery and to prevent the progression into more severe degrees of obstruction. We noticed also that re-stenosis rate was higher in patients who were below two years of age and actually these cases had their peak to peak PG before BPV significantly higher than those above two years (P=0.001), this also was noticed by Ray et al. and McCrindle et al. The subvalvular stenosis of the right ventricle outflow tract was noticed immediately post BPV in 13.6% of patients. These patients received propranolol for 2-3 months period with obvious regression of the subpulmonic obstruction. B-blockers also were used by Fawzy et al., Kassab et al., Moullaert et al., Thapar et al. and Fontes et al., but we can’t draw a conclusion if that regression was due to B-blockers’ effect or was spontaneous due to time, as many studies noticed that the infundibular hypertrophy is reversible in children.

In the current study, over the six year period (range 5 months to 5.2 years) of following up our patients, we noticed also that the residual peak instantaneous gradient had dropped significantly by time, even in those who needed re-intervention. The regression of residual gradient was noticed also by Mahnert et al. over a period shorter than two years in 10 out of 19 cases the PG dropped significantly without any other additional intervention. Also we noticed that the success rate for patients with dysplastic pulmonary valve was 38.8% in comparison with patients with normal pulmonary valve 78% but it was not of statistical significance (P=0.15), but this may be explained by the small number of cases (n=18).

There were studies that indicated the occurrence of pericardial effusion post valvotomy but this was not encountered in our patients. The incidence of pulmonary valve regurgitation increased over the follow up period to 38% and it was near most published series. Possible contributory factors of regurgitation are exacerbation of the anatomic perturbations such as, irregular leaflet tears in the immature valves or avulsions with time. The pulmonary regurgitation was significantly more in cases with higher mean PG pre BPV and those below two years of age. Garty et al. and Rao also noticed that children with small age at the time of dilatation were more likely to develop moderate to severe PR during follow up, but in our cases the pulmonary regurgitation was of mild degree, tolerated by the patients and there was no evidence of right ventricle dilatation or impairment. No immediate or late deaths occurred.

Conclusion
The Balloon pulmonary valvotomy should be the procedure of choice in the treatment of isolated pulmonary stenosis regardless of the severity, because it is safe and effective in treating pulmonary valve stenosis, improving their symptoms and saving the right ventricular function, leaving surgery for those with unsuccessful balloon valvuloplasty. The result of balloon dilatation of dysplastic pulmonary valve was suboptimal. Subpulmonic obstruction post BPV may be regressed spontaneously. The pulmonary regurgitation was of mild degree, tolerated by the patients and there was no evidence of right ventricle dilatation or impairment. Longer-term follow up is needed to
evaluate the significance of pulmonary valve regurgitation.

References


