Introduction

Cardiac malformation with an estimate prevalence of 5 per 1000 live birth is the most common congenital abnormality. Valvular defects are considered the most common cardiac malformation from which 8-12% is related to pulmonary valve stenosis (PVS). It is considered the most benign valvular lesion.

Pulmonary valve stenosis primarily results from a maldevelopment of the pulmonic valve tissue and the distal portion of the bulbuscordis. The stenosis is classified as valvular, subvalvular, or supravalvular. Stenosis may be presented in a typical modification in most of the cases (80-90%) and as the effect of dysplastic changes in 10-20% of cases.

Pulmonic valvar stenosis represents the most frequent cause of right ventricular outflow obstruction, and its related adverse outcomes such as decreased cardiac output, right ventricular hypertrophy, early congestive heart failure (CHF) and cyanosis are directly associated with the right ventricular systolic pressure gradient. Accordingly, it is classified to mild and moderate-to-severe forms. Management of the disease depends on the degree of stenosis.

Over the last few years, balloon pulmonary valvuloplasty have become the treatment of choice for the relief of moderate to severe valvular pulmonic stenosis in neonates, infants, children, and adults. The procedure was first described by Kan et al. It has similar outcomes comparable to surgical valvotomy and is a less invasive, more effective and safe method for relief of right ventricular obstruction in affected children, so surgical pulmonary valvotomy was replaced with this method.

Since then many studies worldwide have investigated the short- and long-term effects of BPV, reduction of the pulmonary valve pressure gradient, normalization of hemodynamic disorders, the need of second intervention, age of treated patients, its complications in patients with PVS and methods for improving the procedure and its outcomes. According to their reports, though the exact indication of BPV is not defined yet but should probably be similar to those used for surgical valvotomy; it is indicated in patients with moderate to severe PVS. The aim of current study was to report our regional survey on the short-term outcomes of BPV among patients with PVS treated in Isfahan Heart center.

Methods

In this cross-sectional study, children with moderate...
and severe pulmonary stenosis aged 0.6-16 years diagnosed in Chamran Hospital, Isfahan-Iran, during 2007-2009 were selected. Those with previous history of surgical valvuloplasty, children with systemic diseases that may affect cardiac function and those that required balloon valvuloplasty as palliative treatment for cyanosis were excluded. Pediatric cardiologist selected the patients who were candidate for BPV. Selected patients underwent cardiac catheterization and angiography and BPV was performed. Right ventricle and pulmonary artery pressure and pressure gradient was measured before and after BPV.

PVS diagnosis and BPV:

PVS diagnosed based on physical examination, ECG, chest radiograph and echocardiographic, hemodynamic and angiographic findings. Before catheterization, the pulmonary artery flow, peak systolic pulmonary artery flow, the size of pulmonary valve annulus for detecting hypoplastic valve, poststenotic dilatation of the pulmonary artery, right ventricular size, wall thickness, and function was measured. The assessments was done using a 3.5-MHz, quantitative, range-gated, two-dimensional, pulsed, echocardiographic Doppler scanner with fast Fourier transform spectral output and a 2.5-MHz phased array with pulsed or continuous-mode Doppler. Color Doppler evaluation in conjunction with 2D echocardiography was used to confirm the clinical diagnosis and quantitating the degree of PVS.

Peak Doppler velocity was used to predict the catheter measured peak to peak gradients across the pulmonary valve. The peak instantaneous Doppler gradient calculated by a modified Bernoulli equation $P = 4V^2$ ($P$: the pressure gradient and $V$: the peak Doppler flow velocity in the main pulmonary artery). The right ventricular peak systolic pressure (RVP) estimated by a modified Bernoulli equation:

$$\text{RVP} = 4V^2 + \text{ERAP}$$

Where $V$ is the peak tricuspid regurgitant jet velocity and ERAP is the estimated right atrial pressure.

Doppler measurements were performed in studied children in resting state and those who were anxious sedated mildly. Different morphology of the pulmonary valve was defined in four categories as follows.

**Typical:** mild to moderately thickened leaflets with evidence of commissural fusion and normal annular dimensions

**Dysplastic:** severely thickened leaflets with evidence of nodular hyperplasia or annular hypoplasia

**Combined:** dysplastic valve morphology as defined but with additional evidence of commissural fusion

**Complex:** other morphologies including primarily post-surgical valvotomy valves and valves involved as part of complex congenital heart lesions

Mild, moderate and severe PVS was defined according to valve gradient as follows.

- **Mild PS:** a valve gradient <35 to 40 mmHg
- **Moderate PS:** the gradient >40 mmHg
- **Severe PS:** a gradient >60 to 70 mmHg

After selecting patients with moderate to severe PVS, hemodynamic cardiac catheterization and angiography was percutaneously performed to confirm the clinical impression and to consider balloon dilatation of the pulmonary valve. Right ventricle and pulmonary artery pressure and peak to peak pressure gradient between RV and PA was measured before and after valvuloplasty.

Studied patients were sedated with intermittent doses of midazolam (0.05 to 0.1 mg/kg IV) and/or fentanyl (0.5 to 1.0 microgram/kg IV) or ketamine before performing balloon pulmonary valvuloplasty. During procedure, ECG monitoring, blood pressure and arterial oxygen saturation measurement was performed by means of a pulse oximeter.

The percutaneous femoral venous route was the most preferred entry site for balloon pulmonary valvuloplasty. A 5 to 7 Fr sheath was inserted into the vein regarding the age and size of the patient as well as the anticipated size of the balloon dilatation catheter. After determining the severity and location of stenosis, an exchange guide wire was introduced through an end-hole catheter and positioned in the distal left pulmonary artery. A balloon that was larger than 20-40% of pulmonary valve annulus size measured by angiography was used and positioned over a guide wire with the valve at its midpoint. As the balloon was inflated, a waist from the stenotic valve was initially observed and disappeared at full inflation (Figure).

Immediately after BPV, reduction of the gradient of transvalvular systolic pressure, reduction of end-diastolic RV pressure, changes of the systolic pressure in the pulmonary artery, presence/intensification of subvalvular stenosis of the pulmonary artery, appearance of insufficiency of the pulmonary valve, or other complications were evaluated.

An effective intervention was defined as one which decreased the stenosis by at least 50% of the initial value. Outcome was classified according to the RV-PA pressure gradient immediately after dilation: Successful if the RV-PA gradient dropped to <36 mmHg and unsuccessful if the RV-PA gradient remained >36 mmHg. Patients were transferred to the intensive care unit until stabilization.

**Statistical Analysis:**

Obtained data were analyzed using SPSS (version 16) software and Student's t-test with 0.05 level of statistical significance was employed. Calculations were made using the STATISTICA statistical program.

**Results**

In this study, 37 patients (11 men and 26 women) were studied. Mean age of studied patients was 4.7 ± 2.8 years with a range of 0.6-16 years. According to echocardiographic findings, typical, dysplastic and complex morphology of the pulmonary valve was presented in 82%, 13% and 5% of patients, respectively. According to echocardiographic
findings, 6 (16%) of patients had moderate PVS and 31 (84%) had severe PVS. Cardiac catheterization findings indicated that mild, moderate and severe PVS was presented in 3 (8.1%), 14 (37.8%) and 20 (54.1%) of patients. Therefore, 34 patients with moderate and severe PVS enrolled.

Among studied population, co-existing cardiac malformations were reported as follows; patent foramen ovale (PFO), small atrial septal defect (ASD), subvalvular pulmonary stenosis (PS), mild aortic stenosis (AS), small ventricular septal defect (VSD), Noonan and hypertrophic cardiomyopathy (HCMP) in 16, 3, 3, 2, 1, 1 and 1 patients, respectively.

A good outcome, defined as a residual catheter gradient ≤36 mmHg, was achieved in 77% of the total group and in 85% of those with typical valve morphology. In contrast, 65% of patients with moderate and severe PVS enrolled. The immediate dilation success rate in our study was 77% (85% in typical forms). The rate was reported to be 88.9% in the study of Liu et al. in China, 81.5% in another study in China, and 90% in the study of Jaing et al. Our findings and the results of previous studies indicated the effectiveness of BPV in the treatment of PVS. The success rate in our study was lower than mentioned ones because 18% of the studied patients had dysplastic or complex morphology. Our results were similar to the findings of Handoka et al. in Egypt with a success rate of 78%, and previous studies showed that factors such as infundibular stenosis and pulmonary valve dysplasia were associated with a lower success rate.

In this study, most of the patients had typical morphology of PVS and those with dysplastic morphology had severe form of PVS, which was similar to those reported by Piotr Werynski et al. in Poland. In their study, 92.4% of patients had typical PVS and those with dysplastic form were associated with moderate to severe forms.

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In this study, mean of RVSP, PAP and the transvalvular peak to peak systolic gradient (ΔP) after intervention decreased significantly in patients with moderate to severe PVS. It was in line with the findings of Jaing et al. in Taiwan, Liu et al. in China, and Piotr Werynski in Poland. Handoka et al. in a similar study among 23 patients with severe PVS reported a significant reduction of the right ventricular systolic pressure, RV-PA systolic pressure gradient and elevation of the pulmonary artery systolic pressure. Moreover, they identified the predictors of successful
pulmonary balloon valvuloplasty and concluded that the immediate post-PBV gradient is the most important predictor in this field.\textsuperscript{20} The balloon diameter to pulmonary valve annulus ratio was 1.28 ± 0.2, which was appropriate for this procedure. Studies in this field indicated that a balloon/annulus ratio of 1.2-1.5 is considered the best for dilatation of the pulmonary valve for appropriate immediate and intermediate outcomes.\textsuperscript{22} Moreover, Rao et al. has recently reported that balloon/annulus ratio of 1.2-1.25 may produce optimal results.\textsuperscript{11} In addition, we obtained a formula regarding the size of balloon and patients age, it would be useful for selection of appropriate balloon based on age.

In current study, we did not observe any complication during and after BPV. Complication rate reported by similar studies was different. Overall, the rate of complication of this procedure is low. According to the report of Valvuloplasty and Angioplasty of Congenital Anomalies (VACA) registry, the rate of complication of BPV was 0.6%, 1.3% and 2.6%, respectively. The complications are as follows; injury of vessel, tricuspid valve, annulus or the cuspid of the pulmonary valve, disruption of the pulmonary trunk, perforation of the RV outflow tract, closure of RV outflow tract, arrhythmias, cardiac tamponade and sudden cardiac arrest.\textsuperscript{23}

The limitation of current study was small sample size of patients and short period of study, i.e. we did not follow up our studied patients for long-term to evaluate the outcome of BPV. In conclusion, BPV was an effective procedure for treatment of PVS in patients with moderate to severe stenosis, as it effectively reduced RV-PA systolic pressure gradient with low complication.

References