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Percutaneous Balloon Valvuloplasty for the Treatment of Pulmonary Valve Stenosis in Children: A Single Centre Experience

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ABSTRACT

Isolated pulmonary valve stenosis (PVS) is a common heart defect (6-9%); the preferred treatment is balloon pulmonary valvuloplasty (BPV). This study aims to assess BPV results in children with isolated PVS treated at the pediatric cardiac catheterization unit, Sohag University Hospital in the period between 2009 and 2012. The studied group consisted of 63 children (37 males and 26 females), aged 29±28 months. The diagnosis of isolated pulmonary stenosis was based on physical examination, ECG, chest radiograph, echocardiography, haemodynamic and angiocardiographic studies. The patients were divided into two groups, depending on successful or unsuccessful procedure. The balloon diameter to pulmonary valve annulus ratio in the two groups was 1.29 ± 0.16, and 1.42 ± 0.13 respectively. Results of our study revealed that patients showed significantly decreased pressure gradient across PVS immediately post-BPV (85.58 ± 15.9 mmHg to 17 ± 13 mmHg) ($p < 0.001$). The procedure was ineffective only in eight (12.7%) children of whom five had re-BPV and three were operated on. Subpulmonary stenosis was seen in 4.8% of children pre-BPV; in 15% post-BPV and in only 3.5% at the end of follow-up. Complications were seen in two (3.2%) patients including one case of a balloon being lodged in the iliac vein (surgical repair) and another patient had perforation of RV during contrast injection with moderate pericardial effusion which resolved spontaneously. Follow-up echocardiography results were similar to those of immediate post BPV values of pressure gradients across PVS. Pulmonary regurgitation > II increased from 2.2% before to 24% after BPV. These medium-term follow-up data confirm efficacy and safety of BPV in children with isolated PVS.

Key words: pulmonary valve stenosis, balloon pulmonary valvuloplasty, pulmonary insufficiency

Introduction

Isolated pulmonary valve stenosis (PVS) is detected in 0.33/1000 newborns and makes up 6-9% of all congenital heart defects among children. Pulmonary valve stenosis appears in a typical modification (80-90%) and as the effect of dysplastic changes (10-20%). The type of stenosis may be the deciding point for the method of therapy, surgical or interventional, and its effects. Since Kan's publication (1982), balloon pulmonary valvuloplasty (BPV) has been the preferred alternative in treatment of the stenosis. The short-term and especially long-term effects of this therapy are still an area of interest of interventional cardiologists in the following aspects: reduction of the transvalvular (pulmonary valve) pressure gradient, normalisation of haemodynamic disorders, the need of second intervention, the occurrence of different complications dependent on type and degree of stenosis, and age of treated patients (Kan *et al.*, 1984, Schmaltz *at al.*, 1989 and Shrivastava *et al.*, 1993). The purpose of this study was to evaluate the early effects of BPV in children with PVS treated at our Pediatric Catheterization Unit, Sohag Faculty of Medicine, Egypt between 2009 and 2012, to assess the results of our newly emerging unit and to update our experience.

Patients and methods:

Patients:

The study population included children with congenital valvular pulmonary stenosis who were subjected to detailed history taking, full clinical examination, resting 12-lead ECG, chest and heart roentgenographic examination and transthoracic echocardiography. Balloon pulmonary valvuloplasty (BPV) was attempted in all children with congenital valvular pulmonary stenosis with a peak-to-peak gradient across the pulmonary valve of 50 mmHg or greater at catheterization-laboratory. The immediate results were classified into two groups based on the right ventricle-pulmonary artery (RV-PA) pressure gradient immediately after dilatation: a successful group in which the RV-PA pressure gradient dropped to <36 mmHg and an unsuccessful group in which the RV-PA pressure gradient was >36 mmHg. All patients were followed up for six months.

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Study Population:

The study included 63 children with isolated congenital valvular pulmonary stenosis recruited from the pediatric cardiology clinic and echocardiography laboratory in Sohag University hospital between May 2009 and April 2012.

Inclusion criteria:

Patients who fulfilled the following criteria were selected for balloon dilatation of the pulmonary valve: Age from birth to 15 years; congenital valvular pulmonary stenosis with peak to peak systolic pressure gradient across the pulmonary valve of >50 mmHg regardless of symptoms.

Exclusion Criteria:

1. Children requiring balloon valvuloplasty as palliative treatment for cyanosis.
2. Children associated with other congenital heart diseases requiring surgical correction.

Methods:

Chest roentgenographs and electrocardiograms were analyzed for chamber enlargement and lung vasculature.

Transthoracic Echocardiography (TTE):

TTE examinations were performed using a Vivid S5 (General Electeronics) with 3 or 5 MHz transducer system. Standard M-mode and 2D-echocardiographic views (parasternal long and short axes views, apical 4, 2 and 5-chamber views, subcostal and suprasternal views) in addition to color Doppler and continuous-wave Doppler were taken to study right ventricular dimensions, pulmonary valve morphology, flow and pulmonary valve annulus. Associated lesions such as PFO, ASD, PDA, post-stenotic dilatation and infundibular stenosis were noted. The maximum peak instantaneous systolic pressure gradient across the pulmonary valve was quantified by the modified Bernoulli equation. Any pulmonary regurgitation was noted and the spatial distribution of pulmonary regurgitant flow was assessed by color flow and pulsed-wave Doppler interrogation. The presence and intensity of pulmonary regurgitation were assessed by color flow mapping. Clinical, standard electrocardiographs (ECG) and 2D Doppler echocardiographic examinations were all performed one day before and after BPV. Complete laboratory evaluation including coagulation profile and kidney function were carried out before catheterization and informed consent was obtained from the parents of each patient.

Procedure:

Premedication with a sedative and analgesic was given to all cases, followed by local anesthesia. Continuous ECG monitoring and blood pressure measurement was carried out as well as the arterial oxygen saturation by means of a pulse oximeter. Before starting catheterization all drugs and equipment for resuscitation were prepared.

The femoral vein was accessed percutaneously and a 5-7F sheath with a diaphragm was introduced into the vein. After heparinization and initial hemodynamic assessment, the pulmonary valve was crossed with an end-hole catheter and pullback was done to document the gradient. An angiographic catheter was then placed in the right ventricular outflow tract and antero-posterior and straight lateral angiograms were performed (1cc/kg over one second) to measure the pulmonary valve annulus and assess the right ventricle and valve anatomy. Lateral projections provided optimal images of the valve hinge point with minimal foreshortening. The angiographic catheter was replaced with an end-hole catheter which was manipulated into the right ventricle with the tip directed toward the right ventricular outflow tract. The pulmonary valve was crossed and the catheter positioned in the left lower lobe branch of the left pulmonary artery. An exchange guidewire was placed in the peripheral pulmonary artery and the catheter replaced with a dilatation balloon. The balloon was centered across the valve annulus in the lateral view and rapidly inflated by hand with diluted contrast material until disappearance of the waist in the balloon. The balloon was then removed while keeping the guidewire in the pulmonary artery. Constant suction on the attached syringe and counter clockwise rotation were important to deliver the deflated balloon. The guidewire permitted repositioning of an end-hole catheter into the distal pulmonary artery to repeat pressure measurements.

The following pressure and hemodynamic measurements were recorded before and immediately after BPV: Right ventricular systolic pressure (RVSP), right ventricular diastolic pressure (RVDP), pulmonary artery

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systolic pressure (PASP), Pulmonary artery diastolic pressure (PAD P), right ventricle-pulmonary (RV-PA) artery systolic pressure gradient and oxygen saturation (SaO_2) assessment by pulse oximeter.

The balloon size was determined according to the maximum internal diameter of the pulmonary valve from hinge point to hinge point during systole measured in the lateral projection of the right ventricle angiogram and corrected for magnification. The balloon diameter to pulmonary valve annulus ratio used in this study was 1.3, and the used balloons were 20mm, 30 mm, 40 mm and 45 mm long.

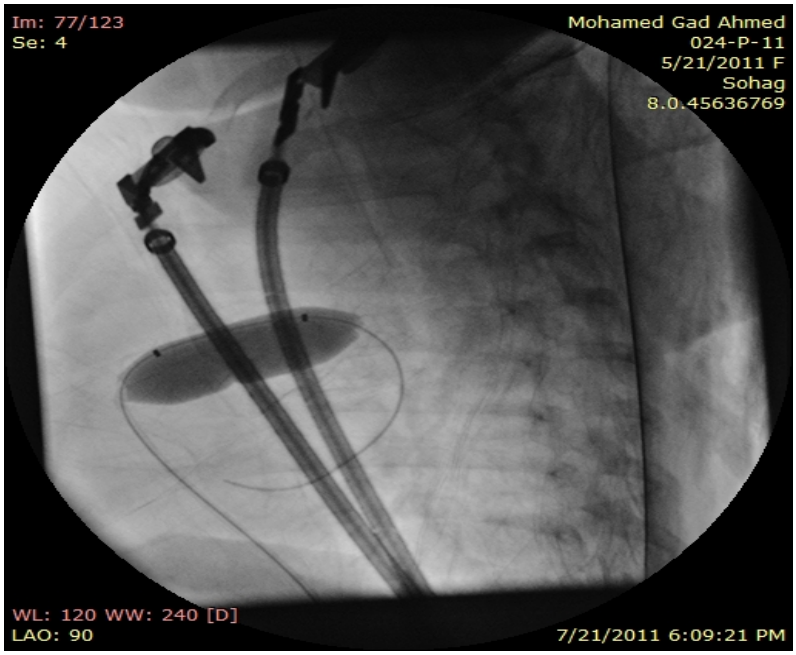

Fig. 1: Balloon valvuloplasty of an infant with severe pulmonary stenosis .Lateral projection shows the inflated balloon with disappearance waist.

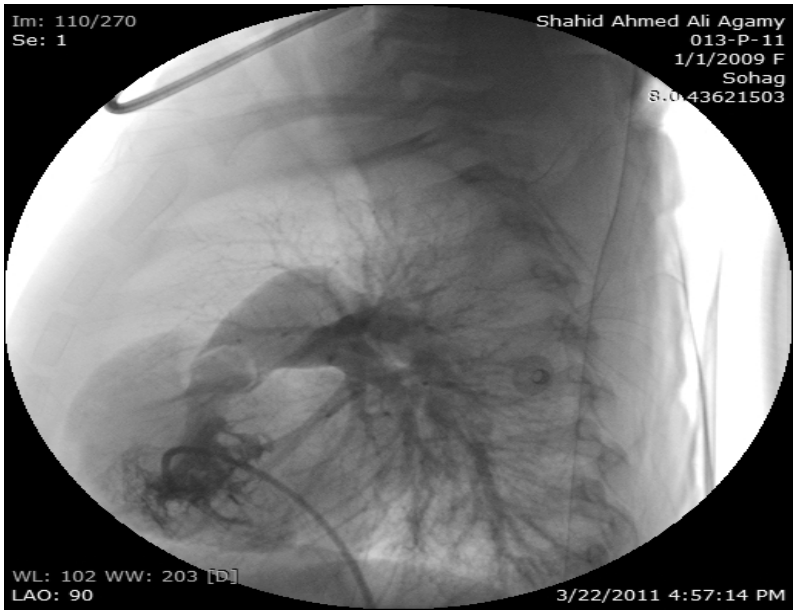

Fig. 2: Angiographic view of the PVS after application of contrast agent to the right ventricle. Domed shape of the valvular cuspids (‘doming)

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Post-Catheterization Care:

Patients were transferred to the intensive care unit where stabilization was carried out together with recording vital signs and pedal pulses. Patients received intravenous fluids, and a broad-spectrum antibiotic (cefotaxime 50mg/kg) was administrated for at least two days.

Follow-Up:

Follow up included clinical evaluation and echocardiographic studies at one month, six months and 1 year. As a considerable number of patients did not complete their follow up visits, we decided to consider the results of follow up till 6 months post BPV. Clinical evaluation focused on symptoms while echocardiography studies assessed the maximum peak instantaneous systolic pressure gradient across the pulmonary valve and the presence of pulmonary regurgitation. 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 Doppler gradient remained >36 mmHg or if the patient required surgery or redilatation after the procedure.

Statistical Analysis:

Continuous variables were expressed as mean ± SD. A paired Student t test was used for comparison of pressure gradients before versus immediately after BPV. Univariate and multivariate stepwise logistic regression analyses were performed to determine predictors of a transpulmonary valve gradient in excess of 35 mmHg. Categorical variables were compared with the use of the chi-square test. *P* value <0.05 was considered statistically significant.

Results:

This study included 63 children with congenital valvular pulmonary stenosis subjected to PBV. The clinical characteristics of the studied patients before PBV are illustrated in (Table 1). The mean age was 29.3 ± 28.1 months; with a male predominance: male/female = (57.7%/ 42.3%); twenty eight children were <12 months; 2 children were neonates; 6 children had Noonan syndrome.

Table 1: Clinical characteristics of patients with valvular pulmonary stenosis.

Variables	Mean ± SD or (%)
Age(months)	29±28.1
Male	58.73%
Female	41.27%
Body weight (kg)	12.21±7.8
Asymptomatic	13(19%)
Dyspnea	29(56%)
Failure to thrive	24(46%)
Recurrent chest infection	12(20%)
Radiological findings	
Normal CTR	*12(22%)*
RVH	*46(88%)*

Table 2: 2D and color-coded Doppler echocardiographic findings of the study population.

Variables	Mean ± SD or (%)
-PV annulus (mm)	10.23 ± 2.2
Peak transvalvular pressure gradient(mmHg)	89.13 ± 18.83
Post-stenotic dilatation	37 (60%)
Infundibular hypertrophy	5 (6%)
Mild pulmonary regurgitation	14 (27%)
PFO	16 (31%)
ASD II	2 (3%)
VSD	3 (4%)
Dextrocardia	1 (2%)

The majority of patients (56%) presented with dyspnea followed by failure to thrive and recurrent chest infections. Cardiomegaly in chest roentography was found in 83%. Evidence of right ventricular hypertrophy in ECG was present in 88% of patients.

From Table (2), the mean baseline peak transvalvular pressure gradient was 89.1±18.8 mmHg. The mean pulmonary valve annulus diameter was 10.9±2.4 mm. Thirty seven patients (60%) had post-stenotic dilatation of the pulmonary artery. PFO was evident in sixteen patients (31%), infundibular hypertrophy present in 5 (6%) patients. One patient had dextrocardia with isolated pulmonary valve stenosis. Mild pulmonary regurgitation was detectable in 14 (27%) patients on initial echocardiographic examination.

One patient had atrial septal defect indicating concomitant closure of defect by Amplatzer septal occulder. Another patient had midmuscular VSD indicating concomitant closure of defect by Amplatzer muscular occluder.

Six months following BPV, the number of patients with pulmonary regurgitation was significantly higher compared to pre-PBV 35 (55.8%) patients. However regurgitation was not more than grade II in severity (73 % grade I and 27 % grade II).

Table 3: The hemodynamic findings before and immediately after pulmonary balloon valvuloplasty

Hemodynamic parameter	Before PBV (Mean±SD)	After PBV (Mean±SD)	*P*
RVSP (mmHg)	97.89±34.38	33.38±7.94	<0.001
SPAP (mmHg)	28.37±14.68	31.28±7.14	<0.001
RV-PA PG (mmHg)	86.2±30.4 mmHg	25.1±16.3 mmHg	<0.001

RVSP: right ventricular systolic pressure, SPAP: systolic pulmonary artery pressure, RV-PA PG: peak-to-peak RV-PA pressure gradient,

Immediately after the procedure patients had significant reduction of RVSP (97.89±34.38 mmHg down to 33.75±7.94mmHg, p<0.001) as well as RVSP-PASP (91 ±30.4 down to 25.1±16.3mmHg, p<0.0001). The pulmonary systolic pressure also exhibited slight elevation post BPV (from 28.37±14.68 mmHg to 31.28± 7.14mmHg, p<0.001) (Table 3).

Table 4: Comparison of the data of immediate successful and unsuccessful pulmonary valvuloplasty

Variable	Successful (n=55) Mean ± SD or (%)	Unsuccessful (n=8) Mean ± SD or (%)	P value
Age	30.54±28.56	19.09±17.83	NS
Gender (M/F)	33(60%)/22(40%)	2(25%)/6(75%)	NS
RVSP before PVBD(mmHg)	87.79±18.38	108.03±22.32	0.007
RVSP immediate PVBD(mmHg)	33.38±7.65	59.43±19.89	0.003
SPAP before BPV	25.17±14.36	24.31±11.31	NS
B/A ratio	1.29 ± 0.16	1.42 ± 0.13	NS
PV annulus(mm)	10.93±2.38	9.01±2.61	NS

RVSP: Rt ventricular systolic pressure, SPAP: systolic pulmonary artery pressure, RV-PA PG: peak-to-peak RV-PA pressure gradient, B/A ratio: balloon to pulmonary annulus ratio.

Patients who did not have a favorable result at 6 months after valvuloplasty had significantly higher RV-PA gradient immediately after PBV compared to the patients who had a favorable result (37±9.80 Vs 10±16.3 mmHg, p<0.001), as well as significant residual elevation of the right ventricular systolic pressure (59.43±19.89 Vs 33.38±7.65 mmHg, p<0.003) .

There was no significant prevalvuloplasty difference in age, gender, RV-PASP before BPV, peak transvalvular PG, balloon/annulus ratio and PV annulus between successful and unsuccessful BPV groups (table 4). The lower immediate post-BPV gradient emerged as the most important predictor of success up to six months post procedure (p<0.001).

In our study, one patients developed major complication in form of perforation of RV during injection of contrast with mild pericardial effusion, child was 3 months and had severe pulmonary valve stenosis and only needed strict observation and procedure postponed after one month. Another severe complication (wedge up of detached balloon in femoral vein) occurred in one patient and needed surgical intervention.

Discussion:

Congenital pulmonary stenosis is a progressive defect, which needs adequate treatment in different periods of life. The decision about the type of treatment should be made according to the degree of haemodynamic changes. Besides very early detection of changes, it is very important to follow them. Even in mild or moderate disease, very fast progression in infancy or early childhood was documented (Kan *et al*., 1982, Kan *et al*., 1984 and Stranger *et al*., 1990). Patients with severe stenosis should undergo treatment even if it is well tolerated and

is asymptomatic at the beginning, because of the possibility of dangerous complications (Schmaltz *et al.,* 1989, Shrivastava *et al.,* 1993). In the present study, successful immediate outcome of BPV was reported in 55 out of 63 cases (87.3%). This rate is in accordance with that of Moura and coworkers (2004). However, at 6 months post BPV the procedure was considered effective in 57 out of the 63 (90.47%) patients. This is explained by regression of the infundibular muscular stenosis during the follow up period in patients with suboptimal immediate results (Masura *et al.,* 1993, Walsh, 2008).

Also Ralf et al.(2012) in multicenter experience found that procedural success was achieved in 91% procedures, being defined as one or more of the following: post-BPV peak systolic valvar gradient to < 25 mm Hg (88%), decrease in gradient by 50% (79%), or reduction of RV/systemic pressure ratio by 50% (45%).

The analysis of our own data showed the mean age of children who underwent the intervention was 29 months. Twenty eight children were less than 12 months. In our study correlations between successful BPV and unsuccessful BPV in relation to balloon to valve hinge point diameter, patient age, associated cardiac lesions, and pre-BPV hemodynamic parameters (RVSP, SPAP, RV-PA PG) were not significantly. This is consistent with the results reported by other studies (Mendelsohn *et al.,* 1996 and Handoka, El-Eraky, 2007). In addition, Mendelsohn and associates observed that the successful BPV group had significantly lower pre BPV gradients and lower systolic pressure ratios than those with unsuccessful results which is consistent to our study.

Balloon size is a determinant factor in achieving good results with percutaneous balloon valvuloplasty. The best results are obtained when balloons larger than the pulmonary annulus are used (Berman *et al.,* 1999). However, in the present study the mean value of balloon/annulus ratio was 1.29 ± 0.16 with no significant difference between successful and unsuccessful groups. This in agreement with the results reported by Mendelsohn et al. (1996) who found that there was no significant difference between successful and unsuccessful groups in terms of age, weight, Z score of the pulmonary annulus or balloon/annulus ratio.

In our study, 5/63 (7.9%) of patients exhibited a rise in the RV-PV gradient at varied time intervals following effective valvuloplasty which needed another BPV. These results are in agreement with those of Rao et al. (1998) who found that 11% of PS patients who underwent BPV had restenosis sometime in the first 2 years post procedure.

Complex echocardiographic diagnosis is the most valuable method in children with PVS. The differences in diameter of the annulus of the pulmonary valve assessed by echocardiography or angiography were insignificant. Currently, the indication for intervention is a transvalvular gradient (pulmonary valve) in Doppler echocardiography > 40 mmHg (Silvilairat *et al.,* 2006). The analysis of echocardiographic data of our patients showed that the majority (87.6%) of children with PVS had the typical form; the dysplastic one was present only in 22.4% and it was associated with moderate and severe stenosis which explained by high percentage of consanguinity of parents in Egypt . Co-existing subvalvular stenosis and insufficiency of the pulmonary valve > II° in Doppler echocardiography was very rare.

Sudden reduction of stenosis of the pulmonary valve leads to increase of the dynamic subvalvular stenosis. This requires the use of propanolol (Walsh, 2003). This relationship was observed in our study, where eleven (17.4%) patients took propanolol, directly after intervention.

Silvilairat *et al.* (2006) also observed that those who obtained incomplete relief of the obstruction immediately after valvuloplasty had a higher baseline PG and higher RVSP which is in agreement to our study.

Despite the experience and technical progress, the valvuloplasty is associated with some risk. The complications are divided into groups: severe, moderate and mild. According to the Valvuloplasty and Angioplasty of Congenital Anomalies (VACA) registry, severe complications are diagnosed in 1.6 % of our series.

The presence or development of mild to moderate pulmonary regurgitation coupled with the lack of significant pulmonary regurgitation due to appropriate B/A ratio without over sizing in our series is consistent with the results of other studies (Hernaez Cobeno *et al.,* 1998, Berman *et al.,* 1999).

The summary data suggest that in long-term observation serious insufficiency is not well tolerated which is not consistent to our study due to lack of long term observation. Shizamaki *et al.* (1984) showed that there were no symptoms at all in 77% of patients for 37 years, 50% of patients for 49 years and only 24% of patients for 64 years. These data are based on analysis of a study on 72 subjects with isolated PVS treated surgically.

Conclusions:

The results of medium-term observations confirm the usefulness and effectiveness of balloon valvuloplasty in the treatment of valvular stenosis of the pulmonary artery even in children with dysplastic changes. Long term follow up (5-10 years) is recommended. The complications of the procedure with increased experience are infrequent.

Study Limitations:

Some limitations exist in the present study. These include the relatively small number of patients and the relatively short period of follow up. More studies with long follow up periods are needed to analyze the important predictors of successful BPV in children and progress of pulmonary insufficiency which is tolerated or not.

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