Iranian Heart Journal; 2016; 17 (2)

Original Article

Ten Years of Experience in a Tertiary Center in Pulmonary Valvuloplasty in Pediatric and Adult Populations

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ABSTRACT

Background: The preferred treatment for isolated pulmonary valve stenosis is percutaneous balloon pulmonary valvuloplasty. The aim of our study was to evaluate the efficacy and short-term results of this procedure in pediatric and adult patients with pulmonary valve stenosis.

Methods: In this retrospective case series, we reviewed the hospital records of pediatric and adult patients with a diagnosis of pulmonary valve stenosis subjected to balloon pulmonary valvuloplasty over a period of 10 years. Data regarding the immediate postprocedural findings as well as echocardiographic transvalvular pressure gradients after 6 months were recorded and analyzed.

Results: Between 2003 and 2013, a total of 248 patients underwent balloon pulmonary valvuloplasty in our institution. Seventy-nine (31.8%) patients were < 18 years of age. The immediate success rate was 61%. However, the drop in right ventricular pulmonary artery pressure gradient was significantly more prevalent in the patients < 18 years old (73.1% in those < 18 y vs. 55.4% in those ≥ 18 y; $P = 0.008$). Regarding the success rate over 6 months after discharge, 75% of the patients < 18 years old and 79% of those ≥18 years old had transvalvular pressure gradients < 50% of the baseline transvalvular pressure gradients on echocardiography performed within 6 months after the initial procedure.

Conclusions: In our case series, we demonstrated that balloon pulmonary valvuloplasty was an effective and safe method for the treatment of pulmonary valve stenosis in both pediatric and adult populations. However, there was a tendency toward a higher postprocedural pressure gradient in the older patients, which made the obstruction more difficult to regress. ***(Iranian Heart Journal 2016; 17(2):30-37)***

Keywords: Congenital heart diseases ■ Pulmonary valve stenosis ■ Balloon valvuloplasty

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Received: November 3, 2015 **Accepted:** April 18, 2016

Isolated pulmonary valve stenosis is one of the most important congenital heart diseases and has an incidence rate of 0.6–0.9 per 1000 live births.[1-3] It is a consequence of the maldevelopment of the distal portion of the bulbus cordis and the pulmonic valve tissue; the valve is thickened and irregular and its annulus may be hypoplastic.[1-3]

Pulmonary valve stenosis is the most common cause of right ventricular (RV) outflow tract obstruction and once left untreated, it results in RV hypertrophy and failure. The severity of RV failure is directly associated with RV systolic pressure gradient. Accordingly, pulmonary valve stenosis is classified as mild, moderate, and severe and its management depends on the degree of stenosis.[1-4]

First introduced by Kan et al., balloon pulmonary valvuloplasty is the preferred method of treatment in these patients. The procedure is feasible and safe, with high success rates in experienced centers.[3-8] Our center is a tertiary center for cardiovascular diseases, including congenital heart diseases, in Iran. This is a report of the short-term results of balloon pulmonary valvuloplasty in pediatric and adult patients with pulmonary valve stenosis over a period of 10 years.

METHODS

In this retrospective study, the hospital records of the pediatric and adult patients who underwent balloon pulmonary valvuloplasty were analyzed. The inclusion criterion was isolated pulmonary valve stenosis, and patients with concomitant congenital anomalies and also those with dysplastic valves were excluded from the study. The demographic and clinical characteristics as well as echocardiographic and catheterization data were recorded from the hospital database.

Among various echocardiographic data, pre- and postprocedural transvalvular pressure gradients, postprocedural pulmonary insufficiency severity, and transvalvular pressure gradients at 1 month's and 6 months' follow-up were collected. From the catheterization reports, pulmonary annulus size, RV systolic pressure, and pulmonary artery pressure before and after the procedure in conjunction with data regarding the procedure—including the methods used for balloon pulmonary valvuloplasty and the balloon size—were obtained.

Considering the pulmonary transvalvular gradient, the severity of the transvalvular pressure gradient was defined as follows[1,9]: mild transvalvular pressure gradient = transvalvular pressure gradient <40mmHg, moderate transvalvular pressure gradient = transvalvular pressure gradient =40–59 mmHg, and severe transvalvular pressure gradient= transvalvular pressure gradient ≥60mmHg.

The severity of pulmonary insufficiency was assessed in accordance with the guidelines of the American Society of Echocardiography.[9]

A successful procedure was defined as a decrease in the transvalvular pressure gradient of at least 50% of the initial value (transvalvular pressure gradient – 50) on the first echocardiography after the procedure (short-term success). A drop in the transvalvular pressure gradient of at least 50% on echocardiography within 6 months after discharge was defined as long-term success.[4, 5, 10-12]

Immediate success was defined as a drop in the RV–pulmonary artery pressure gradient to < 36 mm Hg immediately after dilation. The procedure was defined as unsuccessful if the RV–pulmonary artery gradient remained > 36 mm Hg.[4, 5, 10-12]

The study protocol was approved by the Research and Ethics Committee of Rajaie Cardiovascular, Medical, and Research Center.

Statistical Analysis

The study population was divided into 2 age groups: <18 years old and ≥ 18 years old. The study outcomes were investigated in each group separately. IBM SPSS, version 19 for Windows (IBM Corp., Armonk, NY, USA), was used for the statistical analyses. All variables were assessed in terms of a normal distribution using the Kolmogorov–Smirnov test. The quantitative variables are expressed as means (SDs) or medians (IQRs), as appropriate. The categorical variables are expressed as numbers (percentages). As appropriate, comparisons and associations were tested using the Student *t*-test, the Mann–Whitney test, the χ^2 test, or the Wilcoxon signed-rank test. The multivariable regression analysis was employed to assess the independent predictors of procedural success. A P <0.05 was considered significant.

Table 1. Demographic and echocardiographic findings of both study groups

	<18 y (n=79)	≥18 y (n=169)	*P*
Age (y)	4.4±3.8	25.9±10.8	
Male (%)	53 (67.1)	74 (43.8)	0.001
Dyspnea (%)			
No dyspnea	76(96.2)	30(17.7)	0.02
NYHA class I	0	4(2.4)	
NYHA class II	3(3.8)	135(79.9)	
NYHA class III	0	4(2.4)	
NYHA class IV	0	0	
PS severity (%)			
Mild	10(12.7)	4(2.4)	
Moderate	16(20.3)	27(16)	0.005
Severe	53(67.1)	138(81.7)	
PI severity at baseline (%)			
Non-trivial	38(48.1)	35(20.7)	<0.001
Mild to moderate	40(50.6)	123(72.8)	
Moderate	1(1.2)	11(6.5)	
Moderate to severe	0	0	
Preprocedural PVG (mmHg)	75(50-100)	92(70-130)	0.001

NYHA, New York Heart Association; PI, Pulmonary insufficiency; PS, Pulmonary stenosis; PVG, Pulmonary valve gradient

RESULTS

Totally, 248 patients (52.1% male) were enrolled in this study. The mean of age was 19 ± 13.6 years, with a range of 2.5 months to 68 years. Seventy-nine (32%) patients were < 18 years of age, one-third of whom were infants.

Table 1 depicts the demographic and echocardiographic characteristics of the study population in the 2 age groups.

The most common chief complaint was dyspnea. As is shown in Table 1, the majority of the patients in the group < 18 years of age were asymptomatic and the presence of the RV–pulmonary artery pressure gradient was suspected in the routine pediatric physical examinations. In contrast, our older group was much more symptomatic and, accordingly, both pulmonary valve stenosis and the severity of pulmonary insufficiency were greater in these patients.

Preprocedural Findings and the Procedural Success Rate

The procedure was the single-balloon method in all patients, and the femoral venous route was the preferred site for approach. The mean annulus size was 15 mm (14–20) and 23 mm (23–25), and the mean balloon size was 17 mm (12–20) and 24 mm (24–25) in the group < 18 years old and the group ≥ 18 years of age, respectively.

Regarding the immediate success rate, the RV–pulmonary artery pressure gradient immediately after the procedure dropped to < 36 mm Hg in 61% of the patients, and 79.3% of the patients met the transvalvular pressure gradient – 50 criterion after the procedure.

Table 2 and Table 3 show the pre- and postprocedural hemodynamic data in both patient groups. The 2 population groups of the study exhibited similar results regarding the transvalvular pressure gradient – 50 criterion.

Table 2. Pre- and postprocedural hemodynamic data in the group <18 y

Variables	<18 y (n=79)		*P*
	Preprocedural	postprocedural	
RVSP, mm Hg	92.5(75-120)	44(39.5-60)	<0.001
SPAP, mm Hg	20 (20-25)	20 (20-25)	0.8
PVG, mm Hg			
Baseline	75(50-100)	20(15-40)	<0.001
Within 6 mon		25(16-40)	<0.001[a]
Annulus size, mm	15 (14-20)		
Balloon size, mm	17(12-20)		

a Comparison between PVG at baseline and at 6 months' follow-up
PVG, Pulmonary valve gradient; RVSP, Right ventricular systolic pressure; SPAP, Systolic pulmonary arterial pressure

Table 3. Pre- and postprocedural hemodynamic data in the group ≥ 18 y

Variables	≥18 y (n=169)		*P*
	Preprocedural	Postprocedural	
RVSP, mm Hg	110(85-150)	50 (40-65)	<0.001
SPAP, mm Hg	20(20-24)	20(20-20)	0.8
PVG, mm Hg			
Baseline	92(70-130)	30(20-45)	<0.001
Within 6 mon		30(20-45)	<0.001[a]
Annulus size, mm	23(23-25)		
Balloon size, mm	24(24-25)		

a Comparison between PVG at baseline and at 6 months' follow-up
PVG, Pulmonary valve gradient; RVSP, Right ventricular systolic pressure; SPAP, Systolic pulmonary arterial pressure

Seventy-eight percent in the group < 18 years of age and 79% in the group ≥ 18 years old met the transvalvular pressure gradient – 50 criterion (P = 0.4). However, the drop in the RV–pulmonary artery pressure gradient was significantly more prevalent in the patients < 18 years of age (73.1% in the group < 18 y vs. 55.4% in the group ≥ 18 y; P = 0.008). As regards the long-term success rate, 75% of the patients in the group < 18 years old and 79% of the patients in the group ≥ 18 years of age had transvalvular pressure gradients < 50% of the baseline transvalvular pressure gradient on echocardiography performed within 6 months after the procedure.

The assessment of the patients' symptoms in the first outpatient visit after discharge revealed significant improvement in terms of New York Heart Association (NYHA) class. All patients in the group < 18 years old and 93.8% in the group ≥ 18 years of age were asymptomatic and, consequently, the improvement was significant compared to the preprocedural functional class (P =0.001).

Independent Predictor of a Successful Procedure

To evaluate the independent predictor of a successful procedure, a multivariable analysis using the logistic regression was performed, which demonstrated that the transvalvular pressure gradient before the procedure was an independent predictor of immediate successful balloon pulmonary valvuloplasty (P < 0.001 ; 95% CI: 0.923–0.974).

Complications

Apart from 1 patient with flail pulmonary valve during the procedure, no major peri- or

postprocedural complications were observed in the catheterization laboratory. As was explained, 1 patient (a 23-year-old male) developed partial flail pulmonary valve; he was transferred to the operating room for surgery. The procedure was stopped in only 4 patients before achieving the success criteria: 2 patients had periprocedural arrhythmia causing temporary asystole followed by hypotension and the other 2 had an unsatisfactory immediate result even after 3 balloon inflations.

In both groups, the severity of postprocedural pulmonary insufficiency was mild, but the increase was significant (Table 4 and Table 5). This rise in the severity of pulmonary insufficiency occurred more frequently in the older patients.

Table 4. Comparison of pulmonary insufficiency severity in the group < 18 y

Pulmonary Insufficiency Severity	<18 y (n=79)		*P*
	Preprocedural	postprocedural	
Non-trivial (%)	39(49.4)	35(44.3)	
Mild to moderate (%)	39(49.4)	40(50.6)	
Moderate (%)	1(1.3)	4(5.1)	0.03
Moderate to severe (%)	0	0	

Table 5. Comparison of pulmonary insufficiency severity in the group ≥ 18 y

Pulmonary Insufficiency Severity	≥18 y (n=169)		*P*
	Preprocedural	Postprocedural	
Non-trivial (%)	35(20.7)	29(17.2)	
Mild to moderate (%)	123(72.8)	115(68)	
Moderate (%)	10(5.9)	24(14.2)	0.001
Moderate to severe (%)	1(0.6)	1(0.6)	

DISCUSSION

In the present study, we reported our 10 years of experience vis-à-vis the outcomes of balloon pulmonary valvuloplasty in our institution. We showed that this procedure was safe and highly successful in both pediatric and adult populations. To the best of our knowledge, this study presents the largest series for balloon pulmonary valvuloplasty to date, particularly in adults.

Balloon pulmonary valvuloplasty is the standard method of treatment in patients with pulmonary valve stenosis, with many studies worldwide having investigated its safety and success rate in the short and long terms.[5-7,10-25] In our study, both immediate and short-term success rates were slightly lower than those reported previously—particularly in the adult population. The immediate success rates were 73% and 55.4% in the group < 18 years old and in the group ≥ 18 years of age, respectively. In most studies, the immediate success rate was > 75%–80%.[11,13,14,18,23] In a study by Kaul et al.[18] on 40 patients between 18 and 65 years of age, the immediate success rate was 80%. Jarrar et al.[12] investigated balloon pulmonary valvuloplasty outcomes in 62 patients with pulmonary valve stenosis and showed an immediate success rate of 74%. The study population's age in that study was between 9 months and 44 years old. A success rate > 85% has been reported in pediatric case series.[10,13,23]

It seems that the procedure is more successful in younger patients. Older patients tend to have higher pressure gradients; consequently, earlier intervention at a lower pressure gradient may confer better outcomes.[3,26] In our study, the mean pressure gradient and the severity of pulmonary insufficiency were higher in the group ≥ 18 years of age. This can explain the relatively suboptimal results immediately after the procedure in this group. Fawzy et al.[16] found that the immediate results of balloon pulmonary valvuloplasty might be suboptimal in adults, particularly in those with concomitant infundibular stenosis. The mobility of valve leaflets and the severity of infundibular hypertrophy are the most important predictors of a successful balloon pulmonary valvuloplasty.[26] Our results demonstrated that the preprocedural transvalvular pressure gradient could be an independent predictor of immediate success.

With a decrease in the mobility of the leaflets and an increase in infundibular hypertrophy, the transvalvular pressure gradient increases more and more—which may affect the immediate outcome. On the other hand, it has been shown that the success rate will improve over time. The long-term follow-up of patients who undergo balloon pulmonary valvuloplasty shows that the pressure gradient decreases more after the obstruction is relieved. In patients with severe pulmonary valve stenosis, myocardial hypertrophy results in subvalvular obstruction—which may lead to a suboptimal immediate result. However, this does not necessarily indicate that the procedural outcome is poor because the myocardial hypertrophy may regress after the obstruction is opened and the pressure gradient drops more over time.[18,20,21,24]

In our study, the transvalvular pressure gradient –50 was achieved in 74% of the group < 18 years old and 79% of the group ≥18 years old. Saad et al.[23] and Jarrar et al.[12] showed a progressive reduction in the transvalvular pressure gradient at long-term follow-up.

Another explanation for the relatively suboptimal immediate outcome in our patients could be our relatively lesser expertise in performing balloon pulmonary valvuloplasty in the early years of the study. Be that as it may, we had practically no major peri- and postprocedural complications such as death or cardiac rupture—which is similar to the results reported by most expert centers.

Study Limitations

Firstly, the retrospective nature of the study may have influenced our results. Secondly, we did not report RV size and function and improvement in RV dysfunction following balloon pulmonary valvuloplasty. Thirdly, we did not consider pulmonary valve morphology and infundibular hypertrophy—which may have influenced the success rate. Finally, a short period of follow-up (6 mon) might be considered a limitation.

CONCLUSIONS

Chiming in with previous reports, we showed that balloon pulmonary valvuloplasty was an effective and safe treatment method for pulmonary valve stenosis in both pediatric and adult populations. Nonetheless, there was a tendency toward high pressure gradients in the older patients, which makes the obstruction more difficult to regress. An earlier intervention at a lower gradient may be accompanied by better outcomes and fewer complications. As pulmonary valve stenosis may have an asymptomatic course and present late, particularly in developing countries, educational programs are needed to boost the knowledge of family physicians and pediatricians in this regard.

Financial Support: None.

Conflict of Interest: None.

Ethical Standards: The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant national guidelines on balloon pulmonary valvuloplasty and with the Helsinki Declaration of 1975, as revised in 2008. The study protocol was approved by the Ethics Committee of Rajaie Cardiovascular, Medical, and Research Center.

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