

THE COMPARISON BETWEEN THE ECHOCARDIOGRAPHIC DATA TO THE CARDIAC CATHETERIZATION DATA ON THE DIAGNOSIS, TREATMENT, AND FOLLOW-UP IN PATIENTS DIAGNOSED AS PULMONARY VALVE STENOSIS

DO HOON KIM, MD, SU-JIN PARK, MD, JO WON JUNG, MD, PHD,
NAM KYUN KIM, MD AND JAE YOUNG CHOI, MD, PHD

DIVISION OF PEDIATRIC CARDIOLOGY, SEVERANCE CARDIOVASCULAR HOSPITAL, YONSEI UNIVERSITY HEALTH SYSTEM, SEOUL, KOREA

BACKGROUND: Isolated pulmonary valve stenosis (PS) makes up 6-9% of all congenital heart defects among children. The initial gold standard for diagnosis, follow-up of PS is by echocardiography. However, the most accurate diagnosis still remains to be measurement of the pressure gradient through transcatheterization. The purpose of this study is to compare the difference between the echocardiographic data to the cardiac catheterization data on the diagnosis, treatment, and follow-up in patients diagnosed as PS, and to see what parameters should be closely monitored.

METHODS: A total of 112 patients (Male : Female = 46 : 66) who underwent balloon pulmonary valvuloplasty (BPV) at Severance Cardiovascular Hospital, between December, 2002 to August, 2012 were retrospectively analyzed. The patients were all under 16 years of age and critical PS patients who underwent BPV were excluded from this study.

RESULTS: The pre-BPV right ventricle (RV)-pulmonary artery (PA) systolic pressure gradient and post-BPV systolic pressure gradient showed statistically significant decrease. The pre-BPV RV-PA systolic pressure gradient and 3 month post-BPV systolic pressure gradient showed statistically significant decrease. The consistency between the echocardiographic data and cardiac catheterization data shows statistically significant consistency. The mean pressure gradient and systolic pressure gradient on the echocardiography shows high consistency when comparing with the cardiac catheterization data.

CONCLUSION: Our study shows that BPV in PS is a safe and effective procedure in children and adolescent. The standard echocardiographic evaluation of PS, during diagnosis and follow-up, should include mean transpulmonic pressure gradient, as well as the peak systolic pressure gradient. The success of the procedure should be held off until at least 3 months, only if the patients do not show any symptoms.

KEY WORDS: Pulmonary valve stenosis · Percutaneous balloon pulmonary valvuloplasty · Echocardiography · Systolic pressure gradient · Mean transpulmonic pressure gradient.

INTRODUCTION

Isolated pulmonary valve stenosis (PS) makes up 6-9% of all congenital heart defects among children. PS is divided into valvar, subvalvar, supralvalvar, according to the anatomically stenotic portion, and valvar PS is known to be the most common type. The type of stenosis may be the deciding point for the method of therapy, surgical or interventional, and its effects.¹⁾

PS can be divided into mild, moderate, and severe according to the pressure gradient between the systemic pressure and the right ventricle systolic pressure (RVSP): mild to moderate (RVSP \leq 75% of systemic pressure); severe (RVSP 76-100% of systemic pressure); critical (RVSP > 100%). In the past, these patients were candidates of surgical valvotomy, but in moderate to severe PS, percutaneous balloon pulmonary

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• Address for Correspondence: Jae Young Choi, Division of Pediatric Cardiology, Severance Cardiovascular Hospital, Yonsei University Health System, 50 Yonsei-ro, Seodaemun-gu, Seoul 120-752, Korea Tel: +82-2-2228-8280, Fax: +82-2-393-3080, E-mail: cji0122@yuhs.ac

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valvuloplasty (BPV) has risen as the first treatment option since the first introduction in 1982.²⁾

Since the initial adoption of the procedure, equipment for BPV has improved and the skills of the performers have ameliorated, leading to minimal complications and its usefulness, proven in many previous studies.³⁻⁵⁾

The initial gold standard for diagnosis of PS is by echocardiography. In 2-dimensional (2D) echocardiography there can be evidence of right ventricle (RV) hypertrophy, RV enlargement, or right atrial enlargement. Color flow Doppler imaging demonstrates high-velocity turbulent systolic flow through the pulmonary valve.⁶⁾ Pressure gradients can simultaneously be estimated by continuous wave Doppler.⁷⁾ Pressure gradients consist of echocardiographic systolic pressure gradient and mean pressure gradient. In case of aortic valve stenosis which is similar obstruction disease, mean pressure gradient is considered more important for evaluation of disease among those two gradients.⁸⁾ However, the most accurate diagnosis still remains to be measurement of the pressure gradient through transcatheterization.

The purpose of this study is to compare the difference between the echocardiographic data to the cardiac catheterization data on the diagnosis, treatment, and follow-up in patients diagnosed as PS, and to see what parameters should be closely monitored.

METHODS

SUBJECTS

A total of 112 patients (Male : Female = 46 : 66) who underwent BPV at Severance Cardiovascular Hospital, between December, 2002 to August, 2012 were retrospectively analyzed. The patients were all under 16 years of age and critical PS patients who underwent BPV were excluded from this study. The age range was between 1 month to 192 months and mean age 38.35 months (\pm 48.55 months). Patients with concomitant simple observable heart diseases such as atrial septal defect or patent foramen ovale were included, but those with complex heart diseases were excluded. On retrospective review of the medical files, the main parameters investigated were, age, sex, concomitant diseases, baseline RV and pulmonary artery (PA) pressure gradient measured during catheterization, remaining residual pressure gradient measured during catheterization, pulmonary valve annulus size, balloon size, and minor and major complications.

The type of PS was diagnosed using the 2D echocardiography and the severity was measured using the Doppler echocardiographic measurement assessed by modified Bernoulli equation. Echocardiography datas were collected from randomized results, using our center modality, Philips IE 33 (Philips Andover, Andover, MA, USA), GE vivid E9 (GE Healthcare, Fairfield, CT, USA), The Siemens Acuson sequoia 512 (Siemens Germany, North Rhine-Westphalia, Germany). Parasternal short axis view is mainly used for evaluation of PS,

and additional view was performed, when it was necessary. The peak-to-peak pressure gradient difference between the RV and the PA was measured, as well as the mean transpulmonic pressure gradient. When the gradient on echocardiography was greater than 40 mmHg, or the estimated right ventricular pressure was greater than 50 mmHg, patients who have exertional dyspnea, angina, syncope, or presyncope and RV-PA peak-to-peak gradient greater than 30 mmHg at catheterization, BPV was indicated.⁹⁾¹⁰⁾ During the transcatheter procedure, the pre-procedural measurement between the RV and PA was measured and the post-procedural measurement was also measured, to decide the success of the procedure. We considered the procedure successful when the pressure gradient was less than 20 mmHg.

Follow-up echocardiography was performed within 24 hours of the procedure, 1 week, 3 months, 6 months, and yearly thereafter.

Parametric data were entered into a Microsoft Excel 2007 spreadsheet (Microsoft, Redmond, WA, USA). Results are presented as mean \pm standard deviation or numbers and percentages and analyzed using MedCalc ver 12.3.0.0 (MedCalc Software, Ostend, Belgium).

RESULTS

SUBJECTS

The mean age of the patients were 38.35 months (\pm 48.55 months) and they ranged from 1 month to 192 months (16 years old), the median age was 17 months. Of the 112 total patients, there were 46 male patients (41.07%) and 66 female patients (58.92%).

Patients with concomitant simple observable heart diseases consisted of 32 patients. They were 29 atrial septal defect patients (25.9%), 2 ventricular septal defect patients (1.8%), 3 patent ductus arteriosus patients (2.7%) and 3 patent foramen ovale patients (2.7%).

The mean follow-up duration of the patient was 30.64 months (\pm 26.84 months) ranging from 3 months to 108 months (median, 24 months) (Table 1).

ADVERSE EVENTS

There were no noted major complications. Two cases (1.79%) of minor complications were detected, both of which were spiking fever, but both subsided after a day (Table 1).

ECHOCARDIOGRAPHIC DATA

The type of diagnosed PS were 85 valvular PS (75.9%), 8 supraventricular PS (7%), 17 supraventricular and valvular PS (15.5%), and 2 valvular and subvalvular PS (1.79%) (Table 1). BPV was performed on 8 supraventricular valvular PS patients who showed combined valvular PS on catheterization. The mean pressure gradient measured on pre-BPV echocardiogram between RV-PA was 38.76 mmHg (\pm 16.91 mmHg), ranging from 11 mmHg

to 108 mmHg, and the median was 37 mmHg. The mean pressure gradient measured on echocardiogram within 24 hours post-BPV was 21.33 mmHg (\pm 42.09 mmHg), ranging from 0

mmHg to 73.5 mmHg, and the median was 17.8 mmHg.

The mean pressure difference between the pre-BPV and post-BPV was 17.02 mmHg (\pm 14.39 mmHg), ranging from 0 mmHg to 83 mmHg.

The echocardiographic data of pre-BPV RV-PA systolic pressure gradient, 24 hour post-BPV RV-PA systolic pressure gradient, 1 week post-BPV, and 3 months post-BPV was also analyzed (Fig. 1).

This figure shows the statistical analysis of mean, maximum and minimum results and degree of distribution of variables, indicating the statistical significance of each variables. The pre-BPV RV-PA systolic pressure gradient and post-BPV systolic pressure gradient showed statistically significant decrease, but the difference of 24 hour post-BPV systolic pressure gradient and 1 week post-BPV systolic pressure gradient was not statistically significant. However, the pre-BPV RV-PA systolic pressure gradient and 3 month post-BPV systolic pressure gradient showed statistically significant decrease. On follow-ups, there were 87 patients (77.7%) among 112 patients whose pressure gradient declined over 20 mmHg because of successful procedure, and 25 patients (22.3%) had pressure gradient over 20 mmHg without any symptoms, so the additional procedure was required.

This shows that echocardiography measuring the RV-PA systolic pressure gradient should be regularly followed-up, since the RV-PA systolic pressure gradient shows gradual decrease after the procedure, and the success of the procedure should be determined after at least 3 months, which shows statistically significant decrease in our data.

Table 1. The clinical data of patients with pulmonary valve stenosis

Characteristics	Value (n = 112)
Male, n (%)	46 (41.07)
Female, n (%)	66 (58.92)
Age at procedure (mo)	38.35 \pm 48.55
Follow-up duration (mo)	31.09 \pm 24.0
Complication, n (%)	2 (1.79)
Type of pulmonary stenosis, n (%)	
Valvular	85 (75.9)
Supravalvular	8 (7)
Valvular & supravalvular	17 (15.5)
Valvular & infundibular	2 (1.79)
PV annulus (mm)	13.93 \pm 4.94
Ballon size (mm)	17.5 \pm 5.6
Ballon size/PV annulus	1.27 \pm 0.25

Values are presented as number (%) or mean \pm SD. PV: pulmonary valve

Table 2. The echocardiographic RV-PA systolic pressure gradient during follow-up

Variable (n = 112)	Intraclass correlation coefficient (-1 < ICC \leq 1)
Change in echo data vs. change in cath data	0.69-0.82

Change in echo data: the difference in the RV-PA systolic pressure gradient pre-BPV to RV-PA systolic pressure gradient 24 hour post-BPV on echocardiography, Change in cath data: the difference in the RV-PA systolic pressure gradient pre-BPV to RV-PA systolic pressure gradient post-BPV on cardiac catheterization. ICC: the consistency is higher if the value approaches 1, and the consistency is lower if the value approaches -1, BPV: percutaneous balloon pulmonary valvuloplasty, RV: right ventricle, PA: pulmonary artery

HEMODYNAMIC DATA

The mean RV-PA pressure gradient on cardiac catheterization before BPV was 39.11 mmHg (\pm 16.99 mmHg), the range was between 15 mmHg to 110 mmHg, and the median was 35 mmHg. The mean RV-PA pressure gradient on cardiac catheterization after BPV was 14.14 mmHg (\pm 14.14 mmHg), the range was between 0 mmHg to 60 mmHg, and the median was 10 mmHg.

The mean pressure difference on cardiac catheterization before and after BPV was 24.75 mmHg (\pm 34.77 mmHg), the range was between 2 mmHg to 80 mmHg, and the median was 20 mmHg.

The mean pulmonary valve annulus size was 13.93 mm (\pm 4.94 mm), the range was between 6.5 mm to 31 mm, and the median was 13 mm. The mean balloon size used during the procedure was 17.5 mm (\pm 5.6 mm), the range was between 7 mm to 30 mm, and the median was 16.5 mm. The mean balloon size to pulmonary valve annulus ratio was 1.28 (\pm 0.24), the range was between 0.67 to 2.25, and the median was 1.25.

We compared the difference between the RV-PA systolic pressure gradient before and after BPV on echocardiography against the difference between the RV-PA systolic pressure gradient before and after BPV on cardiac catheterization (Ta-

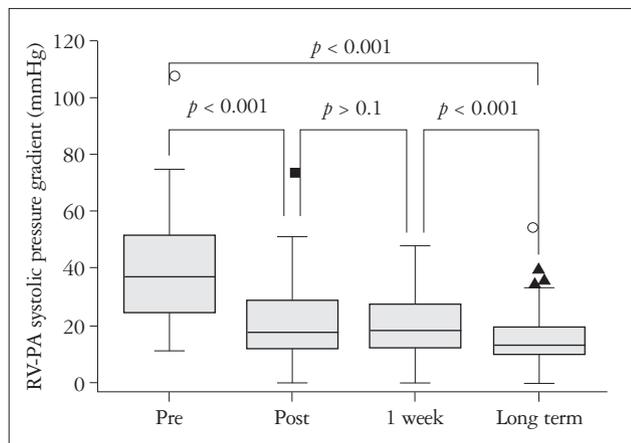


Fig. 1. The echocardiographic RV-PA systolic pressure gradient during follow-up. Pre-BPV RV-PA systolic pressure gradient (pre), 1 day following (post) after BPV, 1 week follow-up (1 week), 3 month follow-up (3 month). Note significant reduction ($p < 0.001$) after BPV which remains unchanged [$p > 0.1$ (0.23)] at 1 week. However, at long term follow-up there was further fall ($p < 0.001$). RV: right ventricle, PA: pulmonary artery, BPV: balloon pulmonary valvuloplasty.

ble 2). Intraclass correlation coefficient was used to compare the consistency of two variables, the measurement is calculated from minimum -1 to maximum 1. As the result gets closer to 1, more consistent measurement can be obtained. If the result is over 0.5, this means the result is the highly consist value. The consistency between the echocardiographic data and cardiac catheterization data was 0.69-0.82, which shows that the decrease in the pressure difference between both data shows statistically significant consistency.

Table 3 and 4 show the consistency between the cardiac catheterization pressure gradient to the echocardiographic systolic pressure gradient, and the cardiac catheterization pressure gradient to the echocardiographic mean transpulmonic pressure gradient. Among 112 patients, 76 patients were enrolled who were able to obtain both parameters. In the pre-BPV data, the intraclass correlation coefficient was 0.79-0.88 in the analysis between the cardiac catheterization data to echocardiographic systolic pressure gradient, which shows relatively higher consistency than the intraclass correlation coefficient (0.55-0.71) between the cardiac catheterization data to echocardiographic mean transpulmonic pressure gradient. However, both values show high consistency, overall.

Table 4 shows the post-BPV analysis. The intraclass correlation coefficient was 0.57-0.73 in the analysis between the cardiac catheterization data to the echocardiographic systolic pressure gradient, which shows relatively lower consistency than the intraclass correlation coefficient (0.61-0.76) between the cardiac catheterization data to echocardiographic mean transpulmonic pressure gradient. However, the values show high consistency, overall. This shows that the mean pressure gradient and systolic pressure gradient on the echocardiography shows high consistency when comparing with the cardiac catheterization data, but the consistency between the mean transpulmonic pressure gradient on the echocardiography was relatively higher after the procedure, which necessitates measurement of mean transpulmonic pressure gradient during echocardiographic follow-up after the procedure.

DISCUSSION

Congenital pulmonary stenosis is a progressive defect, which needs adequate treatment in different periods of life. The decision on the type of treatment should be made, according to the degree of hemodynamic changes and symptoms of the patient. Regular follow-up and very early detection of changes is important. Even in mild or moderate disease very fast progression in infancy or early childhood has been documented.¹¹⁾¹²⁾ 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 complication.¹³⁾¹⁴⁾

Echocardiography plays an essential role in the diagnosis and follow-up of patients with PS. Our main purpose of this study was to see how much the echocardiographic data and the actual hemodynamic data coincide, and to see whether the

Table 3. The comparison between pre-BPV cardiac catheterization data to both echocardiographic RV-PA mean pressure gradient and RV-PA systolic pressure gradient

Variable (n = 76)	Intraclass correlation coefficient (-1 < ICC ≤ 1)
Pre systolic pressure gradient -pre BPV RV-PA pressure gradient	0.79-0.88
Pre mean pressure gradient -pre BPV RV-PA pressure gradient	0.55-0.71

ICC: the consistency is higher if the value approaches 1, and the consistency is lower if the value approaches -1, BPV: percutaneous balloon pulmonary valvuloplasty, RV: right ventricle, PA: pulmonary artery

Table 4. The comparison between post-BPV cardiac catheterization data to both echocardiographic RV-PA mean pressure gradient and RV-PA systolic pressure gradient

Variable (n = 76)	Intraclass correlation coefficient (-1 < ICC ≤ 1)
Post systolic pressure gradient -post BPV RV-PA pressure gradient	0.61-0.76
Post mean pressure gradient -post BPV RV-PA pressure gradient	0.57-0.73

BPV: percutaneous balloon pulmonary valvuloplasty, ICC: the consistency is higher if the value approaches 1, and the consistency is lower if the value approaches -1, RV: right ventricle, PA: pulmonary artery

measured systolic pressure gradient or the mean pressure gradient shows higher consistency. In aortic valve stenosis (AS) patients, the mean transaortic pressure gradient, along with the peak pressure gradient, has taken its place in the standard echocardiographic evaluation. The mean transaortic pressure gradient has been known to show higher consistency than the systolic peak pressure gradient in reflecting the severity, and it has been known to be relatively accurate measurement during follow-up.¹⁵⁾ Because PS and AS fall in the similar obstructive heart disease group, we tried to apply the same concept in PS patients as AS patients.

In this study, the success of BPV was clearly evident in our data, both on the echocardiographic data and the cardiac catheterization data, which shows the efficacy of BPV in the treatment of PS. The pressure decrease was statistically significant, similar to those data that have been proven in many other literatures.²⁾¹⁶⁻¹⁸⁾ Also, in our data, the RV-PA systolic pressure gradient and the mean transpulmonic pressure gradient on the echocardiography showed high consistency to the invasive, but accurate cardiac catheterization data, which indicates that echocardiographic data is a reliable and efficient method in the diagnosis and follow-up of patients with PS, before and after the procedure.

The RV-PA systolic pressure gradient and mean transpulmonic pressure gradient both showed a high consistency to the cardiac catheterization data. However, the pre-BPV data showed relatively higher consistency between the echocardiographic RV-PA systolic pressure gradient to cardiac catheterization data, and the post-BPV data showed relatively higher

consistency between the echocardiographic mean transpulmonic pressure gradient to cardiac catheterization data, which indicates the need to measure both RV-PA systolic pressure gradient and mean transpulmonic pressure gradient, during diagnosis as well as during follow-up.

During the follow-up of patients with PS after BPV, the pressure gradient showed gradual decrease, when comparing the echocardiography performed after 24 hours, 1 week, and 3 months. As long as the patients do not show any noteworthy symptoms, the success of the procedure or the need of re-BPV should be delayed until at least 3 months after the procedure, in the mild to severe PS patients.

In conclusion, our study shows that BPV in PS is a safe and effective procedure in children and adolescent. The standard echocardiographic evaluation of PS, during diagnosis and follow-up, should include mean transpulmonic pressure gradient, as well as the peak systolic pressure gradient. The success of the procedure should be held off until at least 3 months, only if the patients do not show any symptoms.

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