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Long-term follow-up outcomes in patients with pulmonary stenosis monitored at a pediatric cardiology unit

Abstract

Background: Pulmonary Stenosis (PS) refers to various levels of obstruction between the right ventricular outflow tract and the peripheral pulmonary arteries, most commonly occurring at the valvular level. Echocardiography is widely accepted as an effective and reliable tool for the diagnosis and follow-up of PS.

Objectives: This study aims to evaluate the long-term follow-up outcomes of pediatric patients diagnosed with pulmonary stenosis and monitored at a single tertiary center in Turkey.

Methods: We retrospectively analyzed 400 patients with pulmonary stenosis who were followed between 2005 and 2010 at the Pediatric Cardiology Outpatient Clinic of Uludağ University Faculty of Medicine. Diagnosis and follow-up were performed using clinical evaluation and Doppler echocardiography.

Results: Among patients with mild PS, a spontaneous decrease in pressure gradient was observed in 40-50% of cases. In the moderate PS group, 11.4% progressed to severe PS. Across all groups, mean systolic pressure gradients decreased over the follow-up period. The rate of complete resolution in valvular PS decreased with age, with faster improvement observed in children under one year of age. However, progression to severe PS was more frequent in this age group, emphasizing the need for closer follow-up, particularly in infants and those with moderate valvular PS.

Conclusion: Long-term monitoring demonstrates that while spontaneous improvement occurs in many cases, infants and patients with moderate stenosis require closer follow-up due to the higher risk of progression. Early identification and timely intervention remain key to optimal outcomes.

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Introduction

Right ventricular outflow tract obstructions encompass a spectrum of malformations that hinder blood flow from the right ventricle to the pulmonary circulation. These obstructions may occur anywhere from the right ventricular outflow tract to the distal pulmonary arteries and may be isolated or present at multiple levels. Among these, isolated valvular Pulmonary Stenosis (PS) is the most frequently encountered type. Subvalvular and supravalvular forms are less common [1].

Pulmonary stenosis accounts for approximately 7.6% of all congenital heart diseases [2]. In valvular PS, the valve leaflets are thickened, fibrotic, and often fused at the commissures, preventing full opening during systole and leading to elevated right ventricular systolic pressure. This results in a measurable

systolic pressure gradient between the right ventricle and the pulmonary artery. Although cardiac catheterization can provide direct pressure measurements, Doppler echocardiography has become the standard non-invasive modality for assessing the severity of PS [3].

While mild and moderate valvular PS are typically well tolerated clinically, severe cases may require Balloon Valvuloplasty (BVP) or surgical intervention. In the past, surgical valvotomy was the primary treatment [4-6], but since the introduction of percutaneous BVP by Kan et al. [7] in 1982, it has become the preferred initial therapy.

Although several international studies have evaluated the natural history of PS, large-scale data from Turkey remain limited. This study retrospectively investigates the natural course and long-term outcomes of PS in children followed at our center, aiming to contribute to the national literature with one of the largest case series reported from Turkey.

Materials and methods

This retrospective study included 400 pediatric patients diagnosed with Pulmonary Stenosis (PS) who were either followed up in the outpatient Pediatric Cardiology Clinic or hospitalized at Uludağ University Faculty of Medicine between 2005 and 2010. The study was initiated following approval by the Ethics Committee of Uludağ University.

Patient files were retrieved from the hospital archives, and the following clinical and demographic variables were reviewed:

1. Age at diagnosis
2. Etiology of pulmonary stenosis
3. Right Ventricular Systolic Gradient (RVSG) and Ejection Fraction (EF) at diagnosis
4. RVSG and EF during follow-up
5. Presence of Right Ventricular Dysfunction (RVD)
6. Presence of Right Ventricular Hypertrophy (RVH)
7. Post-stenotic dilation at diagnosis or during follow-up
8. Associated valvular regurgitations: Mitral Regurgitation (MR), Aortic Regurgitation (AR), Pulmonary Regurgitation (PR), and Tricuspid Regurgitation (TR)
9. Coexisting congenital cardiac anomalies
10. Symptoms at diagnosis or during follow-up
11. Interventional procedures performed: cardiac catheterization, balloon valvuloplasty, valvulotomy, commissurotomy, and valve replacement
12. Progression or regression of pulmonary stenosis over time
13. Incidence of infective endocarditis according to stenosis severity

Patients were categorized based on their RV systolic pressure gradient as follows:

- Mild PS: RVSG 10-40 mmHg
- Moderate PS: RVSG 40-80 mmHg
- Severe PS: RVSG >80 mmHg

Statistical analysis

All statistical analyses were performed using SPSS software (Statistical Package for the Social Sciences). Descriptive statistics were presented as means ± standard deviations or percentages, as appropriate. A p-value<0.05 was considered statistically significant.

Results

A total of 400 pediatric patients diagnosed with Pulmonary Stenosis (PS) between 2005 and 2010 were included in the study. At the time of diagnosis, 307 patients (76.8%) had mild PS, 44 (11%) had moderate PS, and 49 (12.2%) had severe PS. The mean age at diagnosis was 14.06±27.77 months. There was no significant difference in age at diagnosis among PS severity groups (p=0.47) (Table 1).

Table 1: Frequency and mean age at diagnosis according to severity of Pulmonary Stenosis (PS).

PS severity	Frequency n (%)	Mean age at diagnosis (Months)
Mild PS	307(76.8%)	13.41±26.35
Moderate PS	44(11.0%)	5.76±32.53
Severe PS	49(12.2%)	4.43±5.50
Total	400(100%)	14.06±27.77

PS: Pulmonary Stenosis

Of the 400 patients, 209 (52.2%) were male and 191 (47.8%) were female. There was no statistically significant relationship between PS severity and sex (p 0.05) (Table 2).

Table 2: Distribution of Pulmonary Stenosis (PS) severity by gender.

PS Severity	Male n (%)	Female n (%)
Mild PS	161 (52.4%)	146 (47.6%)
Moderate PS	20 (45.5%)	24 (54.5%)
Severe PS	28 (57.1%)	21 (42.9%)
Total	209 (52.2%)	191 (47.8%)

PS: Pulmonary Stenosis

The distribution of PS types revealed that valvular PS was most common (86.8%), followed by subvalvular (7.2%) and supravalvular (1%) forms. Multiple-level involvement was noted in 5.25% of cases (Table 3).

Table 3: Frequency of Pulmonary Stenosis (PS) types among patients.

PS type	Frequency n (%)
Valvular PS	347 (86.8%)
Subvalvular PS	29 (7.2%)
Supravalvular PS	4 (1.0%)
Valvular + Supravalvular PS	20 (5.0%)
Valvular + Subvalvular PS	1 (0.25%)

PS: Pulmonary Stenosis

Concomitant cardiac anomalies were observed in 55.2% of patients (Table 4).

A statistically significant difference in TR frequency was observed between mild and severe PS groups (p=0.014), but not between other groups (Table 5).

Table 4: Associated cardiac abnormalities in patients with Pulmonary Stenosis (PS).

Associated cardiac condition	Number of patients (%)
None	179 (44.8%)
Mild Aortic Stenosis (AS)	35 (8.8%)
Operated Ventricular Septal Defect (VSD)	41 (10.2%)
Secundum Atrial Septal Defect (ASD)	37 (9.2%)
Ventricular Septal Defect (unoperated)	34 (8.5%)
Patent Foramen Ovale (PFO)	45 (11.2%)
Atrial Septal Aneurysm (ASA)	6 (1.5%)
Mitral Valve Prolapse (MVP)	16 (4.0%)
Operated Atrial Septal Defect	5 (1.2%)
Bicuspid Aortic Valve	2 (0.5%)

PS: Pulmonary Stenosis; AS: Aortic Stenosis; VSD: Ventricular Septal Defect; ASD: Atrial Septal Defect; PFO: Patent Foramen Ovale; IAS: Interatrial Septum.

Table 5: Relationship between PS severity and Tricuspid Regurgitation (TR) at diagnosis.

PS severity	Without TR n (%)	With TR n (%)	Total
Mild PS	42 (13.7%)	265 (86.3%)	307
Moderate PS	5 (11.4%)	39 (88.6%)	44
Severe PS	2 (4.1%)	47 (95.9%)	49
Total	49 (12.2%)	351 (87.8%)	400

PS: Pulmonary Stenosis; TR: Tricuspid Regurgitation

Most patients (79.5%) were referred due to a murmur. Cyanosis, fatigue, tachypnea, and palpitations were less common. There was no statistically significant association between symptom type and PS severity (Tables 6 and 7).

Table 6: Presenting complaints at the time of diagnosis in patients with pulmonary stenosis.

Presenting Complaint	Number of Patients (%)
Asymptomatic	28 (7%)
Murmur	318 (79.5%)
Cyanosis	26 (6.5%)
Easy fatigability	18 (4.5%)
Tachypnea	7 (1.8%)
Palpitations	3 (0.8%)

PS: Pulmonary Stenosis

Table 7: Relationship between presenting complaints and severity of pulmonary stenosis.

PS Severity	Cyanosis	Easy Fatigability	Tachypnea	Palpitations	Asymptomatic	Total
Mild PS	16	10	3	3	275	307
Moderate PS	6	6	2	0	30	44
Severe PS	4	2	2	0	41	49
Total	26	18	7	3	346	400

PS: Pulmonary Stenosis

Right Ventricular Hypertrophy (RVH) and Post-stenotic dilatation

RVH was found in:

- 4.2% of mild PS
- 47.7% of moderate PS
- 61.7% of severe PS

There was a statistically significant difference in RVH prevalence among groups ($p<0.001$). Similarly, post-stenotic dilatation occurred more frequently in moderate and severe PS groups compared to mild PS ($p<0.001$).

Medical and interventional treatment

Most patients (85%) were managed conservatively without intervention. Among those requiring treatment:

- Mild PS: 99% followed medically, 1% received a single Balloon Valvuloplasty (BVP)
- Moderate PS: 50% followed medically, 43.2% received one BVP, and 6.8% received two BVPs
- Severe PS: 67.3% received one BVP, 33.7% received two BVPs

BVP was significantly more frequent in severe PS compared to other groups ($p<0.001$).

Surgical interventions

A minority required surgical treatment: Valvulotomy (2.2%), band resection (1.8%), and displastic valve resection (0.25%).

Progression and Regression of PS

Progression in PS severity was observed in 18 patients (4.5%), all of whom had valvular PS.

- 14 progressed from mild to moderate PS
- 4 progressed from moderate to severe PS

Complete resolution (RVSG<15 mmHg) was observed in 121 patients (30.25%) over a mean follow-up of 48.21±40.86 months.

Regression in severity occurred in:

- 45.6% of mild PS
- 77.2% of moderate PS
- 100% of severe PS

This difference was statistically significant ($p<0.001$; Table 8).

Table 8: Comparison of initial and final systolic pressure gradients in patients with pulmonary stenosis.

	Initial measurement				
Final measurement	10-20 mmHg	21-39 mmHg	40-79 mmHg	>80 mmHg	Total
<10 mmHg	78	39	3	2	122
10-20 mmHg	48	92	21	26	187
21-39 mmHg	10	26	10	21	66
40-79 mmHg	10	4	6	0	21
>80 mmHg	0	0	4	0	4
Total	146	161	44	49	400

Systolic pressure gradients

The overall mean systolic pressure gradient decreased from 35.82 mmHg at diagnosis to 19.09 mmHg at final follow-up (Table 9).

Table 9: Comparison of ınitial and final mean systolic pressure gradients.

PS severity	Number of cases	Initial mean gradient	Final mean gradient
Mild PS	307	23.45 mmHg	16.73 mmHg
Moderate PS	44	58.20 mmHg	25.52 mmHg
Severe PS	49	93.02 mmHg	24.52 mmHg
Total	400	35.82 mmHg	19.09 mmHg

The most pronounced decrease was observed in patients with initially high gradients (moderate to severe PS), with a statistically significant difference ($p<0.001$).

Age and outcome correlation

Improvement rates were inversely proportional to age. Infants, especially those under 1 year of age, had higher rates of spontaneous improvement and greater annual reductions in RVSG. In line with this, the proportion of patients achieving complete resolution was highest in neonates and significantly decreased with age (Table 10).

Table 10: Distribution of cases by age groups and initial doppler echocardiographic pressure gradient measurements.

Age Group	10-20 mmHg	21-39 mmHg	40-79 mmHg	>80 mmHg	Total
0-29 days	45	25	17	10	97
1-5 months	52	58	10	17	137
6-12 months	20	30	8	13	71
13 months-5 yrs	26	31	6	7	70
6-10 years	5	6	2	2	15
>11 years	1	8	1	0	10
Total	149	158	44	49	400

Neonatal subgroup analysis

Among 97 neonates:

- 70 had mild PS, of which 33 completely resolved, 33 remained stable, 4 progressed
- 17 had moderate PS; 1 resolved, 12 regressed to mild, none progressed
- 10 had severe PS; all regressed to mild, none fully resolved (Table 11)

Table 11: Comparison of ınitial and final systolic pressure gradients in newborns with pulmonary stenosis.

	Initial Measurement				
Final Measurement	10-20 mmHg	21-39 mmHg	40-79 mmHg	>80 mmHg	Total
<10 mmHg	26	7	1	0	34
10-20 mmHg	17	14	12	7	50
21-39 mmHg	1	1	4	3	9
40-79 mmHg	0	2	0	0	2
>80 mmHg	1	1	0	0	2
Total	45	25	17	10	97

Discussion

Pulmonary Stenosis (PS) encompasses stenotic lesions occurring at various levels from the right ventricular outflow tract to the peripheral pulmonary arteries, with the most common site of obstruction being the pulmonary valve. Isolated valvular PS is the most frequent type of right ventricular outflow tract obstruction in childhood. In valvular PS, the valve is thickened, fibrotic, the semilunar leaflets are deformed, and commissures are fused. Subvalvular and supravalvular PS are less commonly encountered. In all types, pressure increases proximal to the stenosis and within the right ventricle, leading to right ventricular hypertrophy [3].

In our study, among 400 patients, 347(86.8%) had valvular PS, 29(7.2%) subvalvular PS, and 4(1%) supravalvular PS. Additionally, 20 patients (5%) had a combination of valvular and supravalvular PS.

Echocardiography is a well-established, reliable method for the diagnosis and follow-up of PS. Several studies have demonstrated a strong correlation between Doppler echocardiographic measurements and direct measurements obtained via cardiac catheterization [8,9]. In our study, there were no deaths among the 400 patients (ages 0-18 years) followed for a mean duration of 48.2±40.9 months. This finding aligns with the NHS-2 study by Hayes et al. [10], which indicated that the life expectancy of patients with PS is good and close to that of the general population.

When patients were grouped by the severity of stenosis, 40.7% of those with mild, 88.6% with moderate, and 87.8% with severe PS demonstrated a reduction in pressure gradient over time. Hayes et al. [10] also reported that patients with gradients <25 mmHg had a favorable course. Similarly, Levine et al. [11] found no progression to severe PS in mild cases, and Rowland et al. [12] reported no progression in patients over 2 years old with gradients below 50 mmHg. In another study by Drossner et al. [13], 107 of 146 patients with mild PS experienced complete resolution. Our results support that PS in childhood tends to improve over time, particularly in those with mild or moderate stenosis. More than half of the patients with mild PS achieved complete resolution.

Progression of PS severity was observed in 18 patients (4.5%), all with valvular PS. Among mild cases, 1.3% progressed, whereas 11.4% of moderate cases showed worsening. Drossner et al. [13] reported that three cases with initially mild stenosis progressed to moderate or severe PS. In Rowland et al.'s study [12], four patients under one year old with mild PS progressed to severe PS. Hayes et al. [10] followed 34 patients with moderate valvular stenosis medically and reported that 26(76.5%) progressed to severe PS within one year and underwent valvotomy. In comparison, our progression rates were lower across all groups. Among five patients who progressed to severe PS, two had moderate and three had mild stenosis initially. Hayes et al. [10] noted that most patients progressing to severe PS were initially in the moderate group, which our data also supports. Two of the five patients who progressed to severe PS were under 6 months of age, including one neonate. Drossner et al. [13] similarly reported that three patients progressing to moderate/severe PS were under one year old. Rowland et al. [12] found that 14 of 15 patients who progressed to severe PS were under one year, with 10 of them being neonates. Like previous studies, our findings also suggest that patients under one year are more likely to experience disease progression.

Our study demonstrated that both improvement and progression rates are higher in patients under one year of age. In neonates, physiologic pulmonary hypertension can mask the severity of PS. As this resolves over time, the true severity of the stenosis becomes apparent. This physiologic change may explain why a higher rate of progression to severe PS is seen in this age group. Moreover, the pulmonary valve orifice undergoes significant growth during the first year of life, which can lead to a rapid decrease in systolic pressure gradients and contributes to spontaneous resolution in many patients during infancy.

Hayes et al. [10] suggested that patients with a gradient <25 mmHg are unlikely to experience progression and can be followed annually. Our findings support this recommendation: no progression was observed in patients over 2 years old with a gradient <25 mmHg, and more than half experienced complete resolution.

Rowland et al. [14] recommended follow-up with echocardiography every 6-12 months in patients under 4 years with an average gradient <40 mmHg, and more frequent follow-up in those under 2 years due to higher risk of progression.

Hayes et al. [10] found that the risk of requiring valvotomy during childhood/adolescence was <5% for gradients <25 mmHg, approximately 20% for gradients of 25-49 mmHg, and most patients with gradients of 50-90 mmHg required valvotomy. In our study, valvotomy was performed in 9 patients (2.2%), all with severe valvular PS and gradients >80 mmHg, consistent with previous findings. No intervention was needed for patients with gradients <25 mmHg.

Infective endocarditis is now an uncommon complication of PS. Consistent with the NHS-2 study (10-29), we observed no cases of endocarditis, supporting the low risk of this complication in PS patients.

Subvalvular PS, seen in 7.2% of our patients, is typically caused by infundibular muscular hypertrophy resulting in tubular narrowing below the valve. Supravalvular PS, present in 1% of cases, occurs above the valve and may involve the bifurcation or branch pulmonary arteries. When diffuse in the branches, it is termed peripheral PS and is frequently associated with syndromes such as Alagille, Williams, and congenital rubella. In our cohort, 20 patients (5%) had both valvular and supravalvular PS.

Associated cardiac anomalies included PFO (11.2%), surgically repaired VSD (10.2%), secundum ASD (9.2%), and mild aortic stenosis (8.8%). No additional anomalies were present in 44.8% of patients. The high prevalence of PFO may be related to the younger age at diagnosis, as many patients were diagnosed in infancy.

There was no significant correlation between age at diagnosis and PS severity. While critical PS is often diagnosed in the neonatal period, many children with PS are asymptomatic and diagnosed due to a murmur detected during routine examination. As such, most cases of mild PS are diagnosed early in life, which may explain the lack of a correlation between age at diagnosis and stenosis severity. In fact, 79.5% of our patients were referred for further evaluation after a murmur was detected during examination.

Progressive PS often leads to Right Ventricular Hypertrophy (RVH), dysfunction, and Tricuspid Regurgitation (TR) [3]. In our study, RVH and moderate TR were observed in 47.7% of moderate and 61.7% of severe PS patients. The frequency of RVH and TR was significantly higher in patients with moderate and severe PS compared to mild cases (p=0.012).

Conclusion

This study confirms that valvular pulmonary stenosis is the most common type in childhood and generally follows a benign course. Most cases—particularly mild and moderate PS—show spontaneous improvement or stability over time. Infants under one year require closer follow-up due to both higher improvement and progression rates. Severe PS and progressive cases may require intervention, but overall prognosis remains excellent with appropriate follow-up. Routine echocardiographic monitoring can be safely individualized based on initial gradient and patient age.

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