

## Surgical Management of Infants with Isolated Supravalvular Pulmonary Stenosis: Case Reports

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### ABSTRACT

Pulmonary stenosis (PS) can be seen from the right ventricular outflow tract to the peripheral pulmonary arteries. Most frequently, the obstruction occurs at the level of the pulmonary valve; however, it occurs less frequently at the infundibular level within the trabecular component of the right ventricle or within the pulmonary arterial pathways. Lesions at any of these levels can occur as part of more congenital cardiac malformations such as tetralogy of Fallot, complete transposition of great arteries, or atrial septal defect. Isolated supravalvular pulmonary stenosis (iSPS) is less common than other types of PS. In this study, we present our experience with 4 patients who underwent cardiopulmonary bypass operation for iSPS. In one patient, the circular stenotic area was noted on the touch point of the pulmonary valve. Right ventricular pressures ranged from 70 to 90 mmHg, and the pulmonary artery mean pressures ranged from 14 to 17 mmHg. In all patients, the left ventricular and aortic systolic, diastolic, and mean pressures were moderately increased. Pulmonary artery stenosis was treated successfully using a pericardial or Dacron patch on cardiopulmonary bypass. Various techniques such as balloon dilation have been proposed to deal with this problem, but these may often be unsuccessful because of the elasticity and recoil of the pulmonary artery constrictive ring. Even though endovascular stenting and/or balloon angioplasty have been recently proposed as an initial treatment strategy, they may be associated with some severe complications including pulmonary artery thrombosis or stent migration. Our study, even though it consists of a limited number of cases, suggests that open heart surgery using an oval-shaped patch may be a used as the other main choice for the treatment of iSPS.

### INTRODUCTION

Isolated supravalvular pulmonary stenosis (iSPS) is an uncommon clinical condition compared to other congenital

cardiac diseases, and it is rarely reported in the literature. Stenosis of the pulmonary arterial tree frequently occurs in association with congenital heart diseases such as tetralogy of Fallot or transposition of the great arteries. The stenosis may be localized centrally (within the pulmonary trunk or right and left pulmonary arteries). Four patients, 3 male and 1 female, underwent operation by means of a full median sternotomy for iSPS during the same period. The ages of the patients ranged from 14 months to 4 years. Preoperatively, echocardiographic examinations and cardiac catheterization were performed in all patients. Preoperative echocardiographic and angiographic findings, postoperative right ventricular pressures, and pulmonary artery gradients are summarized in Table 1. The ejection fraction ranged from 65% to 82%, and patients' right ventricular pressures ranged from 70 to 90 mmHg. Pulmonary artery systolic, diastolic, and mean pressures ranged from 10 to 16 mmHg, 18 to 22 mmHg, and 14 to 17 mmHg, respectively. In all patients, the left ventricular and aortic systolic, diastolic, and mean pressures were moderately increased. Preoperative angiographic findings, postoperative pulmonary artery pressures, patients' chromosomal studies, and blood calcium levels are summarized in Table 2.

Three of the 4 patients had perioral cyanosis and diminished exercise tolerance. Midline sternotomy was performed and cardiopulmonary bypass (CPB) was established through aortic and bicaval cannulation with aortic cross clamping. Cardiac arrest was obtained by antegrade delivery of cold crystalloid cardioplegia. The mean pulmonary arteriotomy was performed longitudinally, and the stenotic area was clearly visualized. If there was a circular membrane that caused pulmonary stenosis, it was resected and the pulmonary artery was enlarged using an oval-shaped Dacron or pericardial patch. The average CPB and aortic cross clamp times were  $58 \pm 8$  minutes (range, 50-65 min) and  $45 \pm 8$  minutes (range, 37-55 min), respectively. There were no complications such as low cardiac output, atrial or ventricular arrhythmia, right ventricular failure, or death during the postoperative period. Positive inotropic or chronotropic support was not required for any patient, and digoxin was administered to all patients after the operation. A gradient between the right ventricle and the mean pulmonary artery was found between 10 and 16 mmHg. All patients were discharged home on postoperative day 6 or 8. The patients' serial echocardiographic and physical examinations were normal.

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Table 1. Patient Data

Case No.	Sex	Age at Investigation	Presenting Feature	Second Sound	Ejection Click	Systolic Murmur
1	Male	2 y	Perioral cyanosis, diminished exercise tolerance	Normal	No	Ejection
2	Male	16 mo	Recurrent upper airway infection, body growth retardation	Normal	Yes	Ejection
3	Female	14 mo	Palpitation, diminished exercise tolerance, and perioral cyanosis	Normal	No	Ejection
4	Female	4 y	Accidentally, no cardiac symptom	Normal	Yes	Ejection

**CASE 1**

A 2-year-old boy was admitted to the pediatric cardiology division with perioral cyanosis, a diminished exercise tolerance, and mental retardation. The physical examination revealed that grade 3°/6° systolic murmur could be heard along the left sternal border and that he had chronic liver disease. Routine peripheral blood analysis, electrocardiography (ECG), and chest x-ray film were normal. He had undergone an unsuccessful pulmonary angioplasty for main pulmonary artery stenosis (PAS) 6 months before at another institution. iSPS was seen and a 70 mmHg systolic gradient was detected across the supravalvular pulmonary area by means of transthoracic echocardiography. Left and right heart catheterizations were planned. Systolic, diastolic, and mean pulmonary artery pressures were recorded as 18, 2, and 9 mmHg, respectively, and systolic pressures of 70, 151, and 132 mmHg were recorded across the right ventricle, left ventricle, and the aorta, respectively. The diaphragma-like iSPS was seen in the lateral position of the right heart catheterization (Figure 1 and Figure 2). He underwent operation and the stenotic segment of the main pulmonary artery was enlarged with an oval-shaped Dacron patch that was sutured with a 6/0 polypropylene suture. In the postoperative period, echocardiography did not reveal any significant gradient on the pulmonary artery. He was discharged from the hospital on postoperative day 7. Six months after the operation, serial echocardiographic examinations were normal.

**CASE 2**

A 16-month-old boy was referred to our institution and presented with a recurrent upper airway infection and mental and physical retardation. In the physical examination, a grade 3° to 4°/6° systolic murmur was heard along the left sternal border. Routine peripheral blood analysis, ECG, and chest x-ray film were normal. There was no chromosomal anomaly in his genetic research. iSPS was noted and transthoracic echocardiography recorded a 70-mmHg systolic gradient across the supravalvular pulmonary area. Left and right heart catheterizations were performed. Systolic, diastolic, and mean pulmonary artery pressures were recorded as 25, 8, and 17 mmHg, respectively. Systolic pressures of 65, 110, and 80 mmHg were recorded across the right ventricle, left ventricle, and the aorta, respectively. The diaphragma-like iSPS was seen in the lateral position of the right heart catheterization. He underwent operation and the stenotic segment of the main pulmonary artery was enlarged with an oval-shaped Dacron patch that was sutured with a 6/0 polypropylene suture. A 10-mmHg systolic gradient was noted on the pulmonary artery. He was discharged home with good clinical results. Three and 6 months after the operation, serial echocardiographic examinations were normal.

**CASE 3**

A 14-month-old girl was brought to our hospital for evaluation of palpitation, diminished exercise tolerance, and peri-

Table 2. Operative Data\*

Case No.	RVH	LVH	Perioperative PAP, mmHg	Perioperative Mean PAP, mmHg	RVP, mmHg	Aortic/LV, mmHG	Additional Cardiac Anomalies	Chromosomal Studies	Serum Calcium	Perioperative PA Systolic Gradient	Postoperative	
											PA Systolic Gradient, mmHG	Surgical Procedure
Case 1	Yes	No	18/2; Mean, 9	12	70	151/132	No	Normal	Normal	70	15	Oval patching with Dacron
Case 2	Yes	No	25/8; Mean, 17	15	65	110/80	PFO	Normal	Normal	84	10	Oval patching with Dacron
Case 3	Yes	Moderate	22/11; Mean, 16	17	90	96/126	PFO	Normal	Normal	70	11	Oval patching with Dacron
Case 4	No	No	18/10; Mean, 14	13	60	115/120	No	Normal	Normal	80	16	Oval patching with Dacron

\*RVH indicates right ventricular hypertrophy; LVH, left ventricular hypertrophy; PAP, pulmonary artery pressure; LV, left ventricle; PA, pulmonary artery; PFO, patent foramen ovale.

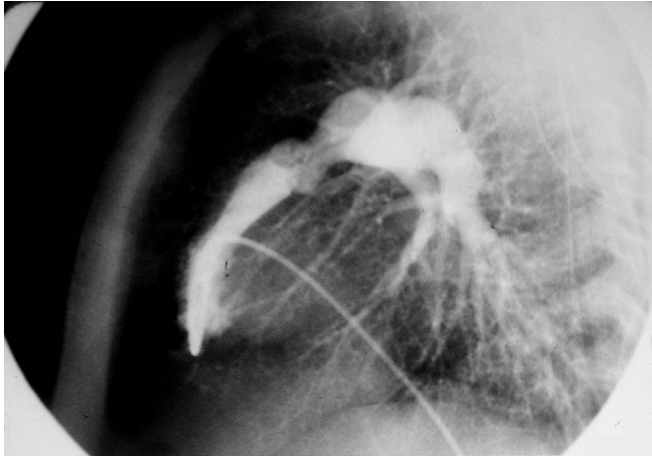


Figure 1. First angiogram of Case 1, doughnut viewed from the side appearance in the area between the pulmonary valve and supralvalvular diaphragm (lateral and postero-anterior position).

oral cyanosis developing during crying. Routine blood analysis, ECG, and chest x-ray film were normal. A systolic murmur was heard on the left parasternal border. Transthoracic echocardiography and cardiac catheterization demonstrated supralvalvular pulmonary stenosis. The left ventricular end diastolic and end systolic pressures were 20 mmHg and 11.6 mmHg, respectively. In addition, there was a supralvalvular stenotic area developing on the touch point of the pulmonary valve in this patient (Figures 3-5; the serial echocardiographic findings are presented in Figure 6). She also had a patent foramen ovale. A 70-mmHg systolic gradient was found between the right ventricle and part of the supralvalvular pulmonary area. Surgical correction was carried out using the standard CPB technique. Supralvalvular stenosis was repaired with a pericardial patch and the patent foramen ovale was closed. Pulmonary artery gradient diminished to 12 to 14 mmHg. She was in good health with normal exercise tolerance. She was discharged from the hospital on the sixth day. Six months after the operation, physical and transtho-

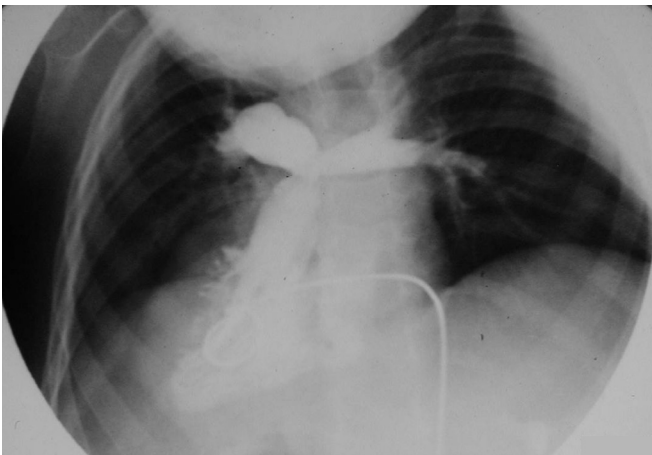


Figure 2. Second angiogram of Case 1, doughnut viewed from the side appearance in the area between the pulmonary valve and supralvalvular diaphragm (lateral and postero-anterior position).



Figure 3. Close view of the pulmonary valve. Two white arrows indicate the stenotic area. PV indicates pulmonary valve.

racic echocardiographic examination and the performance status were normal.

#### CASE 4

A 4-year-old girl without any symptoms was referred from another hospital to our Pediatric Cardiology unit because she was suspected of having a congenital heart defect. The physical examination revealed only that a grade 2° to 3°/6° systolic murmur could be heard along the left sternal border. Routine blood analysis, ECG, and chest x-ray film were normal. Supralvalvular pulmonary artery stenosis was noted and a



Figure 4. The touch point of the stenotic pulmonary area when the pulmonary valve is totally opened (black arrow).

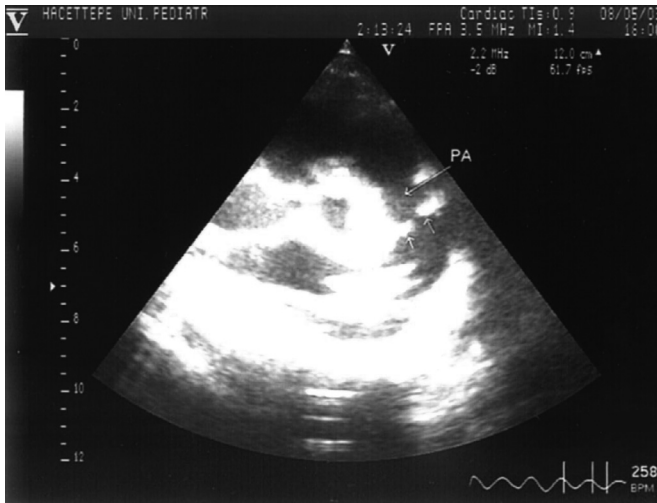


Figure 5. The circular type supravalvular pulmonary artery (PA) stenosis is seen in the transthoracic echocardiographic examination (white arrows).

60-mmHg systolic gradient was detected across the supravalvular area of the main pulmonary artery by means of transthoracic echocardiography, and left and right heart catheterizations were performed. Systolic, diastolic, and mean pulmonary artery pressures were recorded as 18, 10, and 14 mmHg, respectively. Systolic pressures of 60, 115, and 120 mmHg were recorded across the right ventricle, left ventricle, and the aorta, respectively. An iSPS was seen in the lateral position of the right heart catheterization. In the operation, a segment of the stenotic main pulmonary artery was enlarged with an oval-shaped Dacron patch that was sutured with a 6/0 polypropylene suture. In the postoperative period, echocardiography did not reveal any significant gradient on the pulmonary artery. She was discharged from the hospital on postoperative day 6. Serial echocardiographic examinations revealed cardiac functions were normal, and the supravalvular pulmonary gradient disappeared.

## DISCUSSION

PAS can be seen from the right ventricular outflow tract to the peripheral pulmonary arteries. Most frequently, the obstruction occurs at the level of the pulmonary valve; it occurs less frequently at the infundibular level within the trabecular component of the right ventricle or within the pulmonary arterial pathways. The typical cardiovascular findings in iSPS are a normal first heart sound, a systolic ejection click, and an ejection systolic murmur.

Only a few reports concerning iSPS were found in our research of the literature. In 1967, Schlesinger and Meester reported 14 patients with iSPS, and Roberts and colleagues later reviewed 15 patients with iSPS [Schlesinger 1967; Roberts 1973]. Although isolated stenosis of the pulmonary arteries can also occur in various parts of the pulmonary arterial tree and is frequently associated with congenital rubella [Rowe 1963], Williams syndrome [Williams 1961], or Alagille syndrome [Alagille 1975], antenatal histories of our



Figure 6. Transthoracic echocardiograms of Case 3. This figure clearly shows the pulmonary valvular touch point in the supravalvular stenotic area in the systolic and diastolic period. In the second echocardiogram, the 2 short arrows indicate the stenosis in the supravalvular area; A and B indicates infra and supravalvular pulmonary areas in systol of the right ventricle. The pulmonary valve is touching near the supravalvular circular ring. The other echocardiograms display the systolo-diastolic relationship between the pulmonary valve and the circular stenotic area. PA indicates pulmonary artery.

patients did not reveal any evidence of infection or drug ingestion. Hastraider et al reported that the pulmonary vasculature was invariably involved in the congenital Rubella syndrome. There was a thin supravalvular diaphragm with post-stenotic dilatation on the angiographic examination [Hastraider 1967].

On the other hand, stenosis of the pulmonary artery is frequently associated with complex congenital heart diseases such as tetralogy of Fallot, complete transposition of great vessels, or supravalvular aortic stenosis. One of our patients had patent foramen ovale and no signs of physical appearance of Williams or Aegille syndrome. Two patients had mental retardation.

We did not perform any histopathologic examination from the resected pulmonary tissue, but medial elastic tissue with intimal proliferation has been defined as an etiologic mechanism by Rowe et al in their reported cases [Rowe 1978].

The typical indicator of iSPS is an elevated right ventricular pressure in the presence of normal systemic arterial pressure. According to the literature, and as occurred in our cases, right ventricular and proximal pulmonary arterial pressures are elevated. The pulmonary arterial systolic pressure rose before the pulmonary valve was crossed. When the systolic pressure falls below 60 mmHg it is considered a mild stenosis. The degree of right ventricular hypertension is the main indicator of severity. All of our patients had severely increased right ventricular pressure and 3 of the 4 were symptomatic. The cardiothoracic ratio was normal in all patients except for one who had mild right ventricular enlargement apparent in the chest x-ray films. In every patient, the right ventricular angiogram showed a constant segmental area or narrowing, simulating a diaphragm in the main pulmonary artery, approximately 1 to 1.5 cm distal to the pulmonary valve. Interestingly, a supravalvular stenotic area was recorded on the touch point of the pulmonary valve in one patient. This finding leads us to possibly conclude that this chronic valvular traumatic effect may be the cause of iSPS development. There was no evidence of hypertrophic cardiomyopathy, subvalvular PAS, or poststenotic pulmonary artery dilatation in any patient. The aortic valve and supra-aortic area were normal.

Surgical or interventional alternatives such as percutaneous balloon angioplasty and/or stenting are suggested for the relief of the pulmonary artery and/or its branch stenosis. The success rate of this treatment plays a central role in the long-term outcomes [Arnold 1988; Rothman 1990; Hosking 1992; Castenada 1994; Fogelman 1995]. However, the success rate and the rate of complications due to angioplasty procedure in the Hosking study were reported as 53% of cases and 5% of cases, respectively [Hosking 1992]. On the other hand, stents are rarely used in infants because stents do not grow as the patient grows [Castenada 1994]. Pulmonary artery dilation using a percutaneous balloon sometimes requires the use of endovascular stents and fails because of marked vessel compliance, which usually leads to kinking or folding of the vascular channel. Although the safety of catheter dilation of pulmonary arteries has improved with experience, morbidity persists and rare deaths continue to occur during the process of balloon dilation because the

intima and media begin to tear before the pulmonary artery enlarges [Hosking 1992; Castenada 1994]. Prevention of recoil allows stents to dilate the pulmonary artery branches more effectively than balloon dilation alone. But stents may promote intimal hyperplasia in pulmonary arteries, causing late stenosis, and do not grow as the child grows. In addition, it is hard to position them accurately in a vascular tree with short segment vessels [Castenada 1994], and the proximity of the pulmonary valve prohibits the use of a stent. As a result, we preferred open heart surgery in the described iSPS cases. In the long-term follow-up, no symptoms were observed and there was no evidence of residual gradient in the echocardiographic examinations.

The surgical approach prevents the intimal or medial tearing, and, if necessary, the surgeon can resect the pathologic area and perform an end-to-end main pulmonary artery anastomosis safely under CPB. Recently, Bacha et al have reported a different technique for the repair of iSPS without using the patch [Bacha 2004]. Findings and the technical approach for the surgical management of stenosis of the branch pulmonary artery has been published by McGoon and Kincaid [1964]. This described technique proposed for selected cases requires extensive pulmonary mobilizing. Therefore, we recommend using our surgical approach in particular cases such as these because it can be performed easily and does not need extensive pulmonary mobilization. In each case, a vertical incision was made through the narrowed area and was sutured horizontally with a rectangular Dacron or pericardial patch. The remaining gradient was found on the pulmonary artery between 10 to 16 mmHg.

Increasing right ventricular hypertrophy on the ECG or echocardiogram suggests that the patient has entered the severe category and relief should not be delayed. Balloon angioplasty was the first treatment undertaken, as reported by Hosking et al [1992]. They reported good immediate results in about half of their patients; however, restenosis developed in 17% of their cases. On the other hand, Fogelman and colleagues have reported successful results with the use of endovascular balloon-expandable stents in 42 patients [Fogelman 1995]. However, 29 cases needed recatheterization and various degrees of acquired intraluminal narrowing were documented in their study. In the same study, 15 stents required redilation because of intraluminal stent obstruction. Furthermore, the chance of complications due to percutaneous dilation may increase during the intervention. Also, the clinical follow-up for these cases was too short.

In conclusion, we suggest that enlargement of the supravalvular PAS with a pericardial or Dacron patch is safe and technically easy under CPB and has provided good results during chronic follow-up in infants with iSPS; however, successful treatment is achieved by balloon dilation for accompanying pulmonary artery branch stenosis. Unsuccessful results recognized after CPB can be corrected by recreation of the patching using the same operation.

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## REVIEW AND COMMENTARY

### *Editorial Review Board Comment 1:*

Please clarify the conclusion in the abstract, particularly regarding the touch point of opening of the pulmonary valve.

### *Author's Response by Dr. Dogan:*

In one case, the supravalvular stenotic area was recorded at the end of the pulmonary valves in each right ventricular systol. In our opinion, although this conclusion is speculative, chronic valvular mechanical stress might be a cause of developing or increasing intravascular endothelial pathology due to the chronic intimal damage or stress. To date, this or a similar mechanism has not been reported as a cause of PAS. This hypothesis may not be explained with our single defined case because histopathologic examination from the resected pulmonary tissue was not performed in this patient. Nevertheless, medial elastic tissue together with intimal proliferation has been defined as an etiologic mechanism by Rowe [1978]. We therefore suggested the relationship between the pulmonary valves and the pulmonary endovascular pathology with this case.

### *Editorial Review Board Comment 2:*

In the discussion, the authors mention papers on the same topic. The results in the current study should be compared with these publications. Were the results concerning uncomplicated conduct of the operation and good clinical outcome the same?

### *Author's Response by Dr. Dogan:*

Isolated supravalvular PAS is an extremely rare clinical condition. A number of studies about the use of a percutaneous approach for peripheral PAS are found in the medical literature; unfortunately, there is only a limited similar study about the described topic in the literature. To compare our study with a similar one and its outcomes, another report of such surgical experiences is needed. However, we could not found any similar study in the English literature.

### *Editorial Review Board Comment 3:*

A discussion on the type of patch material used in these operations would be interesting.

### *Author's Response by Dr. Dogan:*

There is no common consensus about the use of patch material in these particular groups. Recently, 8 cases that were treated surgically using autologous repair and underwent CPB have been reported by Bacha et al [Bacha 2004]. In these cases, a different surgical approach without the use of external patching was reported. However, this technique needed extensive pulmonary mobilizing and its CPB time was longer than that of our surgical approach. As such, Bacha's technical approach may be used in selected cases but our surgical approach of using a simple patching technique is easily performed without any complication.

Traditionally in our institution, we use a patch of Dacron material for the creation of a right ventricular outflow tract in cases with tetralogy of Fallot or isolated stenotic right ventricular outflow tract obstruction. There is no complication in our cases' clinical follow-up after the use of synthetic material. As a result, in our opinion, both the pericardial and the synthetic patch may be used to treat this pathology because these 2 patch materials (glutaraldehyde fixed pericard) are easily manipulated and sutured.

### *Editorial Review Board Comment 4:*

The results in the Hosking study should be specified. Why were long-term results of balloon angioplasty disappointing?

### *Author's Response by Dr. Dogan:*

Currently, balloon angioplasty and/or intravascular pulmonary artery stenting have been increasingly used in cases with main branch or peripheral pulmonary artery stenoses. Successful results using percutaneous arterioplasty or stenting have been reported by Hosking et al and Fogelman et al in their respective studies [Hosking 1992; Fogelman 1995]. Although initial treatment choice is different from institution to institution, we believe that successful results will be obtained in the near future with the help of an experienced cardiology unit. Hosking et al reported a successful outcome in 53% of their cases, and recurrent pulmonary stenoses developed in 17% of their cases. On the other hand, although 1 catheterization was performed in their study, the remaining 16 cases required reintervention or recatheterization. Some complications such as femoral venous thromboses, pulmonary artery laceration, or aneurysm of the pulmonary

artery may be seen after these procedures. Indeed, they reported in the same article that 5% of the procedures were associated with complications. In this study, balloon arterioplasty was performed for peripheral pulmonary stenoses in a number of the cases; however, there was a recoiling effect reported in 2 of their cases with discrete supra-valvular stenosis after a le Compte maneuver for the correction of transposition of great arteries. Stenting of the pulmonary artery (this procedure also requires an experienced cardiologist) has been used in selected pediatric groups [Fogelman 1995]. However, reduced flow to the branch pulmonary artery due to the straddling of the stents in 28% of cases was reported. Also, distal pulmonary artery thrombosis and protrusion into the pulmonary artery was reported by the authors as a severe complication. The proximity of the pulmonary valve prohibits the use of a stent. Although the follow-up time in the Fogelman study was too short, the above studies suggest perfect outcomes in the early and midterm period. We believe that balloon angioplasty and/or stenting may be used as an effective and initial choice of method for pulmonary branch

stenoses, and the recoiling effect and restenoses in the early period should be properly considered by the interventional cardiologist. As such, the surgical approach may be the main solution for this group of patients. Percutaneous procedure is an effective treatment in cases with branch pulmonary artery stenoses, especially for peripheral and multiple stenoses, and we believe that open heart surgery can be performed easily and reliably with successful results.

*Editorial Review Board Comment 5:*

In the Conclusion, the authors state that the remaining gradient on the pulmonary artery was within "acceptable limits," but above they say in all cases echocardiographic examinations were normal.

*Author's Response by Dr. Dogan:*

Preoperatively, in all 4 patients, the pulmonary artery systolic gradient was noted between 70 and 84 mmHg. In the early period, the echocardiographic examinations revealed that the patients' pulmonary artery gradient was recorded between 10 or 16 mmHg. Therefore, these limits were found to be within the normal or acceptable limits.