

# Surgical Correction of Congenital Aortic and Pulmonary Stenosis in an Adult

## Erişkinde Konjenital Aort ve Pulmoner Stenozu Cerrahi Onarımı

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### ABSTRACT

Although aortic and pulmonary valve stenosis are among the most common congenital heart defects, the combination of both aortic and pulmonary valve stenosis in the same patient appears to be very uncommon. Accurate diagnosis of combined valvular stenosis is imperative prior to surgical correction otherwise surgery of only one of the lesions may result in an insufficient hemodynamic improvement. A 34-year-old man with congenital aortic and pulmonary valve stenosis underwent Nick's operation, aortic valve replacement, pulmonary valvotomy and resection of anomalous muscle of the right ventricular outflow tract. The operation was successfully performed and postoperative course was uneventful.

**Key Words:** Aortic valve stenosis; pulmonary valve stenosis; ventricular outflow obstruction; adult; congenital.

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### ÖZET

Aort ve pulmoner kapak darlığı en sık görülen konjenital kalp anomalileri olmasına rağmen aynı hastada kombine olarak görülmesi nadir görülen bir durumdur. Mevcut lezyonların sadece birisine cerrahi uygulaması, hemodinamik düzelmenin yetersiz kalmasıyla sonuçlanabileceğinden, cerrahi onarımı öncesi kombine valvular stenozun kesin tanısının konulması zorunludur. Konjenital aort darlığı ile seyreden pulmoner stenozu olan 34 yaşında erkek hastaya Nick operasyonu, aort kapak replasmanı, pulmoner valvotomi ve sağ ventrikül çıkım yolundaki anormal kas bandı rezeksiyonu uygulandı. Ameliyat başarıyla gerçekleşmiş olup, ameliyat sonrası dönemi sorunsuz seyretti.

**Anahtar Kelimeler:** Aort kapak darlığı; pulmoner kapak darlığı; ventrikül çıkım engeli; yetişkin; konjenital.

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## INTRODUCTION

Although aortic and pulmonary valvular stenosis are among the most common congenital heart defects, the combination of both aortic and pulmonary valvular stenosis in the same patient appears to be very uncommon. The purpose of this report is to describe a successful surgical treatment of congenital combined valvular aortic and pulmonary stenosis in an adult.

## CASE REPORT

A 34-year-old man presented to our hospital with dyspnea and fatigue. His exercise tolerance has always been limited. Physical examination was normal except for 4/6 systolic murmurs on pulmonary and aortic areas. Chest X-ray film disclosed the heart and pulmonary vessels to be of normal size, and electrocardiogram (ECG) showed biventricular hypertrophy. Transthoracic echocardiography (TTE) was performed, and biventricular hypertrophy and a stenotic bicuspid aortic valve with a mean gradient of 68 mmHg were noted (Figure 1). Although right ventricular hypertrophy was present, stenosis in the right ventricular outflow (RVOT) could not be evaluated fully because of suboptimal TTE images.

Transesophageal echocardiography (TEE) was subsequently performed, and there was valvular aortic stenosis (gradient: 114/68 mmHg), valvular pulmonary stenosis (gradient: 93/56 mmHg), minimal aortic regurgitation and severe biventricular hypertrophy. 64-slice computed tomography was done for detailed evaluation of RVOT and pulmonary artery, and a severe stenosis of pulmonary infundibulum and poststenotic dilatation was observed. Moreover, aortic root was found to be smaller than the arch of aorta, and measured to be 2 cm at the sinotubular junction (Figure 2).

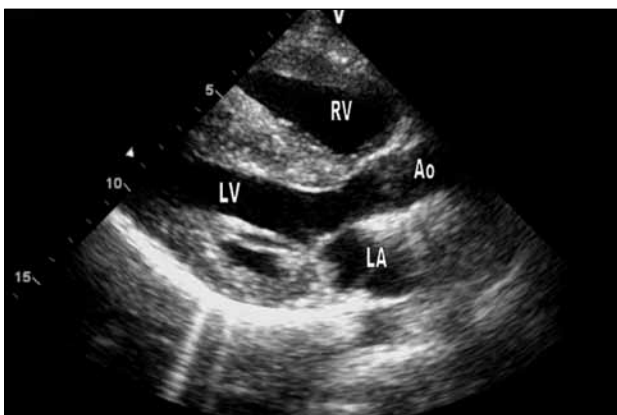


Figure 1. The image of transthoracic echocardiography.

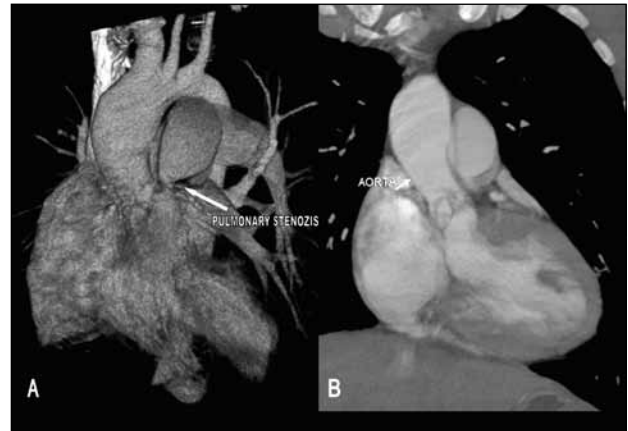


Figure 2. The images of 64-slice computed tomography.

On catheterization, a peak-to-peak gradient of 70 mmHg was found at the level of pulmonary infundibulum. Surgical operation was planned for the symptomatic severe aortic stenosis and pulmonary stenosis. At operation, the pericardial space was found to be obliterated by fibrous adhesions. A thrill was palpated in the ascending aorta. After cannulation and cross clamp were performed, aortotomy was done. The bicuspid and heavily calcified aortic valve was resected. There was obstruction in LVOT and it was enlarged by Nick's procedure. Then, we implanted No. 21 prosthetic aortic valve (Figure 3). Right ventriculotomy was made for exploration of pulmonary valve. The pulmonary valve was dome-shaped, had a small central orifice, and was free of gross calcific deposits. But the infundibular area was hypertrophic, and muscular resection and repair with a pericardial patch were performed (Figure 4). Measurement of pressure af-

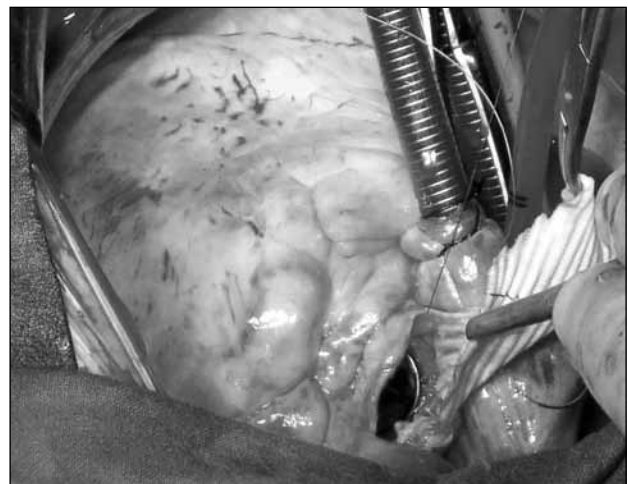
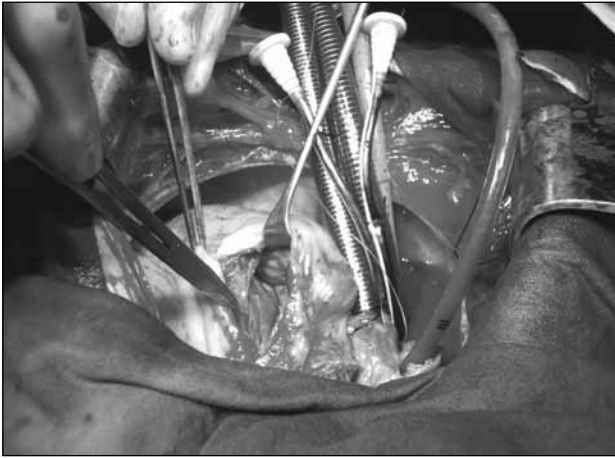


Figure 3. Obstruction in LVOT was enlarged by Nick's procedure.



**Figure 4.** The infundibular area was hypertrophic.

ter the operation showed a 7 mmHg peak systolic pressure gradient between right ventricular body and pulmonary trunk and no peak systolic pressure gradient between left ventricle and aorta. The post-operative follow-up was uneventful, and he was discharged one week later.

## DISCUSSION

With the development of cardiac surgery, the combined anomaly reported is potentially correctable, and successful surgical correction has been reported. Congenital valvular aortic, subaortic stenosis and infundibular or subvalvular pulmonary stenosis are relatively common congenital cardiac anomalies when they occur as isolated defects or in association with other intracardiac deformities<sup>(1)</sup>. Shemin et al., described two patient with combined valvular pulmonary and aortic stenosis associated with a fossa ovale type atrial septal defect<sup>(2)</sup>.

Presenting symptoms of combined aortic and pulmonary stenosis depend on how severely and quickly the condition develops. Symptoms include are dyspnea, shortness of breath, fainting with physical exertion, chest pain and fatigue. Milazzo et al. reported an infant with subvalvar and valvar pulmonary stenosis, subvalvar, and valvar aortic stenosis and hypertrophic cardiomyopathy, who presented with pulmonary hemorrhage<sup>(3)</sup>.

The combined aortic and pulmonary stenosis can be suspected from the clinical examination. If the clinical features suggest pulmonary stenosis and there is a left ventricular heave, aortic stenosis should be suspected. If a thrill is present on both the left and right second intercostal spaces or if the systolic murmur is heard almost equally well at both areas, a combined lesion should be considered.

Detection of radiation of murmurs to both the upper back, as in pulmonary stenosis and to the carotids, as in aortic stenosis might be helpful<sup>(4)</sup>. Our patient had murmur at both pulmonary and aortic areas.

The diagnosis of congenital pulmonary stenosis, valvular and infundibular, and aortic stenosis appear to be substantiated by the conformity between clinical features and the results of the specific tests such as TEE, catheterization, and 64-slice computed tomography<sup>(5)</sup>.

For reconstruction of LVOT there are a number of procedures, namely, Nick, Manugian, Konno and Raston. As our patient's aort annulus was not very narrow, we used Nick's procedure to reconstruct LVOT. After that we replaced the aortic valve because it was congenitally bicuspid, heavily calcified and not suitable for reconstruction.

It seems of great importance that a surgical plan for this condition provide for relief of both obstructions at the same operation. If the aortic valve alone is solved, severe postural hypotension and death are likely. After correction of the infundibular pulmonary stenosis the pulmonary flow may increase with a greater volume of blood delivered to the left ventricle. If obstruction of the left ventricle outflow is not relieved, the ventricle may fail, causing fatal pulmonary congestion. Thus the diagnosis of combined valvular lesions, when clinically suspected, should be confirmed by catheterization prior to any planned surgery<sup>(6-8)</sup>.

Percutaneous ballon aortic valvuloplasty is now treatment option for congenital valvular aortic stenosis except possibly in the elderly. Pulmonary ballon valvuloplasty has been extensively utilized for relief of pulmonary valve stenosis<sup>(9,10)</sup>. Percutaneous ballon aortic valvuloplasty was not suitable in our patient because aortic valve is bicuspid and heavily calcified.

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