

# Peripheral Pulmonary Artery Stenosis in Adults

## Yetişkinlerde Periferik Pulmoner Arterin Stenozu

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**ABSTRACT** Congenital left or right pulmonary artery stenosis is a rare anomaly, which usually coexists with other cardiac anomalies. In adulthood, only 1/16 of pulmonary artery stenosis is isolated. Stenosis may be single or multiple. Pulmonary angiography has been accepted as the gold standard diagnostic tool, however, the pulmonary arterial anatomy can be delineated better by newer techniques such as three dimensional reconstruction of computed tomographic images. In this paper, we reported two cases of different type congenital stenosis of pulmonary artery, in which a simple echocardiographic finding helped us to suspect the stenosis. The diagnosis was confirmed by pulmonary angiography and computed tomography.

**Key Words:** Echocardiography, cardiac catheterization,  
computed tomography, pulmonary artery

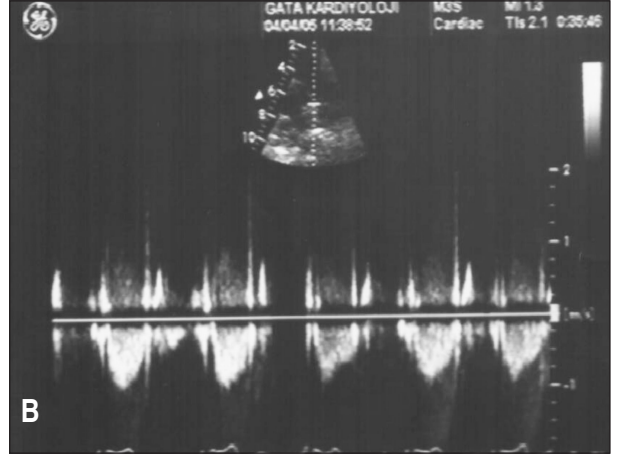
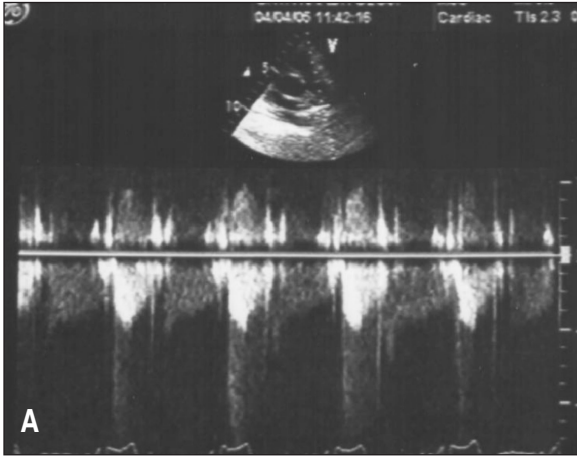
**ÖZET** Konjenital sol veya sağ pulmoner arter darlığı, genellikle başka kardiyak anomalilerle birlikte bulunan nadir bir anomalidir. Erişkin çağda pulmoner arter darlıklarının 1/16'sı izoledir. Darlık tek veya birden çok olabilir. Tanıda altın standard pulmoner anjiyografidir, ancak günümüzde bilgisayarlı tomografi ile elde edilen üç boyutlu rekonstrüksiyon gibi daha yeni tekniklerle anatomi daha iyi ortaya konulabilmektedir. Bu yazımızda basit ekokardiyografik bulgulardan şüphelenerek ortaya koyduğumuz konjenital pulmoner arter darlığının iki değişik tipini sunuyoruz. Tanı pulmoner anjiyografi ve bilgisayarlı tomografi ile de teyit edilmiştir.

**Anahtar Kelimeler:** Ekokardiyografi, kardiyak kateterizasyon,  
bilgisayarlı tomografi, pulmoner arter

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**P**ulmonary artery stenosis is the narrowing of the pulmonary trunk, bifurcation or branches. It was first described in 1938, and classified by Oram in 1964.<sup>1</sup> However, Gay's classification<sup>2</sup> of supralvalvular pulmonary stenosis into 4 types is more widely used. Etiology is not well-known, but coexistence with other congenital heart disease is well documented.<sup>2,3</sup> In clinical practice, various symptoms may manifest depending on the severity of stenosis and coexisting cardiac and pulmonary disorders. Some cases may mimic pulmonary embolism and cause misdiagnosis.

The classification of Gay and co-worker, who divided pulmonary artery stenosis in 4 types, is commonly accepted. Type I involves a single stenosis of the main, right or left pulmonary arteries, without peripheral branch involvement. Type II involves stenosis at the bifurcation of the pulmonary trunk extending into the origins of the right and left pulmonary arteries,



**FIGURE 1:** A; Transthoracic echocardiography revealed 32 mm Hg pressure gradient with continuous Doppler beam placed on left pulmonary artery, and B; when the Doppler beam placed on right pulmonary artery the pressure gradient was 8 mm Hg.

without peripheral branch involvement. Type III involves multiple stenosis of the peripheral pulmonary branches, without central involvement. Type IV involves combination of the central and peripheral pulmonary artery stenosis.

Here we report two cases of peripheral pulmonary artery stenosis suspected upon transthoracic echocardiography. Written informed consents were obtained from both patients to publish their medical material for scientific purposes.

### CASE 1

A 24 year-old male with dyspnea and syncope was admitted to our clinic. On physical examination, crescendo-decrescendo 3/6 systolic murmur was heard centrally in the vicinity of the second left intercostal space, radiating to the left axilla. On his transthoracic echocardiography, 2D findings were in normal range. Continuous Doppler interrogation of pulmonary flow revealed two different gradients depending on the line of the ultrasonic beam; when the beam included the left pulmonary artery, the pressure gradient was 32 mmHg (Figure 1A) while it was 8 mmHg when it included only the right pulmonary artery (Figure 1B). His three-dimensional reconstruction of multi-detector computed tomographic images showed coarctation and distal narrowing of left pulmonary artery (Figure 2). On his pulmonary artery catheterization, there was 32 mmHg peak gradient (mean 21 mmHg) across the distal segments of left pulmonary artery and bifur-

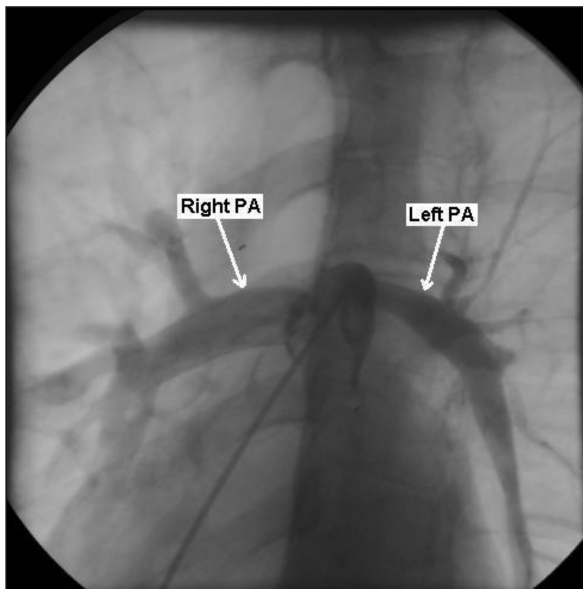


**FIGURE 2:** Three-dimensional reconstruction of multidetector computed tomographic view of pulmonary arteries. Right PA: right pulmonary artery; Left PA: left pulmonary artery.

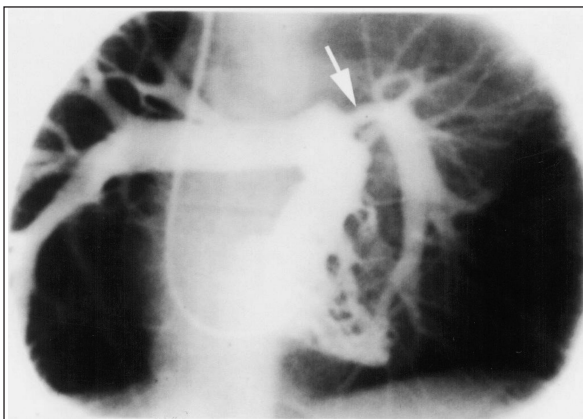
cation; with the pressure of right ventricle 57/31/10 mmHg, pulmonary truncus 50/24/13 mmHg, and left pulmonary branch 15/10/5 mmHg. During pull-back of the catheter, we recorded all the segments of left pulmonary artery for any other narrowing. According to the Gay's classification, this was type IV (Figure 3).

### CASE 2

A 31 year-old male was diagnosed as chronic pulmonary embolism and pulmonary infarction and has been followed-up for 10 years in other centers. His complaint progressed and patient was admitted to our hospital because of bloody sputum and dys-



**FIGURE 3:** Angiographic view of pulmonary artery. Note the peripheral narrowing of left pulmonary artery.



**FIGURE 4:** Angiographic view of pulmonary artery. Arrow indicates left pulmonary artery narrowing.

plea. The physical examination revealed late systolic murmur on the second intercostal area. His transthoracic echocardiography revealed a peak gradient of 23 mmHg (mean 11 mmHg) on pulmonary trunk. On the pulmonary artery catheterization, there was 37 mmHg gradient, (mean gradient 17 mmHg) between proximal and distal segments of the left pulmonary artery. The pulmonary perfusion-ventilation scintigraphy revealed a left pulmonary fixed defect. Pulmonary stenosis was differentiated from pulmonary infarction by pulmonary catheterization (Figure 4).

## DISCUSSION

Pulmonary artery coarctation is a rarely reported entity in adulthood.<sup>4</sup> Most of the cases are diagnosed during infancy because of clinical incompetence. Among the reported concomitant cardiac abnormalities, aortic supravalvular stenosis, aortic hypoplasia, aortic coarctation, mitral valve prolapse, septal defects, tetralogy of Fallot, major aortopulmonary collateral artery and renal artery stenosis can be listed. Sugayama et al<sup>5</sup> reported that only one out of 16 cases was isolated. Iatrogenic supravalvular pulmonary stenosis after repair of Fallot tetralogy has also been reported.<sup>6</sup>

Catheterization is the gold standard for diagnosis, determination of pressure gradient across the coarctation and poststenotic dilatation. The clinical picture or echocardiographic findings generally resembles to that of valvular pulmonary stenosis. In our first case, a suspicious TTE finding was found. The difference in the peak velocity of blood in the left and right pulmonary artery was more than that can be explained by different  $\theta$  angles. For this reason, the stenosis was suspected in the left pulmonary artery and was confirmed by further examination. On the other hand, this finding was not observed in our second case. The only explanation is that the pulmonary coarctation in the Case 2 was localized more distally than the Case 1.

Asymmetric lung circulation may cause serious disturbance of the ventilatory/perfusion ratio on nuclear imaging. This is a confusing finding because the patients with this finding may be misdiagnosed as idiopathic and recurrent pulmonary embolism, which was the situation as in the case of 2. Absence of any coagulation abnormality suggests that the stenosis is not due to thrombus.

During the physical examination of patients with childhood congenital pulmonary stenosis, there may be additional findings about the etiology, such as chubby round bloated face of infant with typical mobile dome-shaped pulmonary valve stenosis, facial appearance of arteriohepatic dysplasia (Alagille syndrome); deeply set eyes, prominent overhanging forehead and pointed chin, webbing of the neck, low-set ears, abnormal air-

clesand hypertelorism (Noonan syndrome); mental retardation, abnormal facial appearance, supraaortic stenosis with hypercalcemia (Williams syndrome); hypermobility of soft tissue (Ehler-Danlos syndrome); and congenital cataract and skin findings (congenital Rubella syndrome). Both of our cases were free of above findings. According to Gay's classification, Case 1 was classified as type IV and the Case 2 as type I.

In patients with isolated left pulmonary artery stenosis where flow rate to the lung is less than

30% of right ventricular output balloon dilatation with or without stent insertion is effective in relieving the obstruction. In other patients with more diffuse pulmonary artery stenosis, such as bilateral involvement, intervention is generally restricted to the cases having systemic or suprasystemic right ventricular pressure levels.<sup>7</sup>

In conclusion; TTE may provide invaluable information about pulmonary artery stenosis as in our cases, so that it can be very helpful in planning invasive procedures.

## REFERENCES

1. Oram S, Pattinson N, Davies P. Post-valvular stenosis of the pulmonary artery and its branches. *Br Heart J* 1964;26:832-47.
2. Gay BB Jr, French RH, Shuford WH, Rogers JV Jr. The roentgenologic features of single and multiple coarctations of the pulmonary artery and branches. *Am J Roentgenol Radium Ther Nucl Med* 1963;90:599-613.
3. Elzenga NJ, von Suylen RJ, Frohn-Mulder I, Essed CE, Bos E, Quaegebeur JM. Juxtaductal pulmonary artery coarctation. An underestimated cause of branch pulmonary artery stenosis in patients with pulmonary atresia or stenosis and a ventricular septal defect. *J Thorac Cardiovasc Surg* 1990;100:416-24.
4. Kreutzer J, Landzberg MJ, Preminger TJ, Mandell VS, Treves ST, Reid LM, et al. Isolated peripheral pulmonary artery stenoses in the adult. *Circulation* 1996;93:1417-23.
5. Sugayama SM, Moisés RL, Wagênfur J, Ikari NM, Abe KT, Leone C, et al. Williams-Beuren syndrome: cardiovascular abnormalities in 20 patients diagnosed with fluorescence in situ hybridization. *Arq Bras Cardiol* 2003;81:462-73.
6. Kirklin JW, Appelbaum A, Barger LM Jr. Primary repair versus banding for ventricular septal defects in infants. In: Kidd BSL, Rowe RD, eds. *The Child with Congenital Heart Disease after Surgery*. Mount Kisco New York: Futura; 1976.p.3-5.
7. Webb GD, Smallhorn JF, Therrien J, Redington AN. Diseases of the heart, pericardium, and pulmonary vascular bed. In: Zipes DP, Libby P, Bonow RO, Braunwald E, eds. *Braunwald's heart disease a textbook of cardiovascular medicine*. 7th ed. Philadelphia: Elsevier Saunders; 2005. p.1542.