

## Combined congenital coronary artery anomaly: dual left anterior descending coronary artery and absence of left circumflex artery

*Kombine konjenital koroner anomalisi: Çift sol ön inen koroner arter ve sirkumfleks arter yokluğu*

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### Introduction

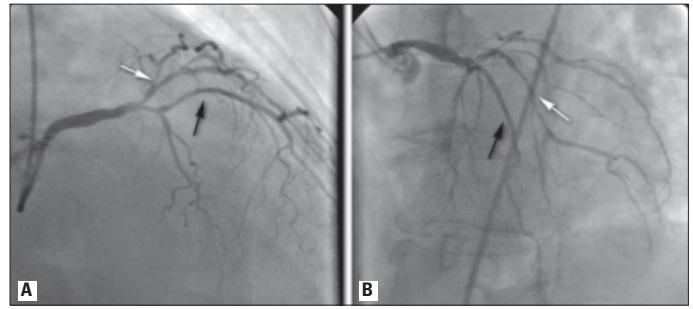
Because coronary artery anomalies are usually asymptomatic, their incidence is not exactly known in normal population. However, the putative incidence of coronary anomalies was between 0.6-1.3 percent in general population, which had been estimated in coronary angiography series (1, 2). The coincidence of two coronary anomalies is uncommon. In this report, we defined combined coronary anomaly, which consisted of both dual left anterior descending (LAD) coronary artery and absence of left circumflex (Cx) artery in the same patient. The frequency of the absence of the left circumflex artery is reported as 0.003% (4/126.595 cases) in coronary angiography studies. In this case, the super dominance of the right coronary artery (RCA) is seen. Right coronary artery (RCA) super dominance associated with various syndromes has been reported (2, 3). The combination of these two rare coronary anomalies is unusual. We could not find any reports in the literature describing the coincidence of these two coronary anomalies.

### Case Report

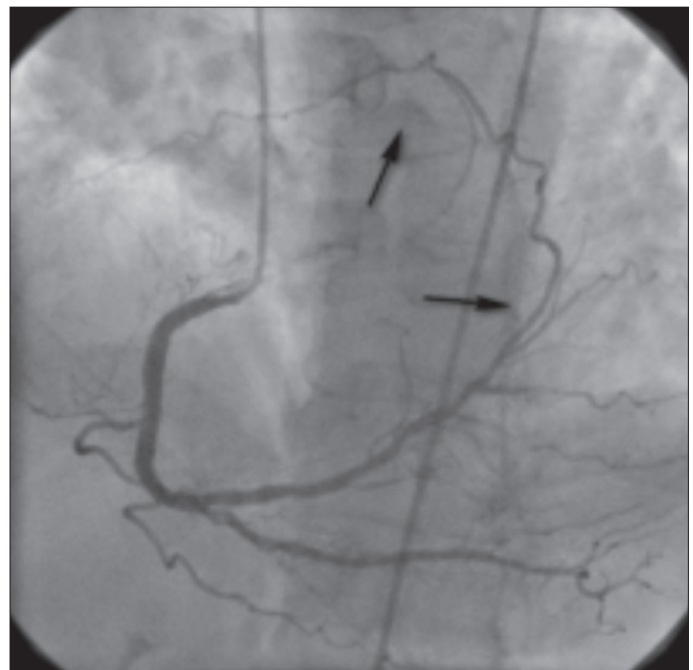
A 67-year-old female patient was presented with a typical exertion of angina pectoris. Our patient had cardiovascular risk factors of hypertension, old age, and menopause. We decided to make coronary angiography due to positive exercise stress test. Electrocardiogram revealed inverted T waves in leads V2-6. Echocardiography showed (no regional wall motion abnormality) normal wall motion and ejection fraction (65%). In coronary angiography, in the region matching to the LAD artery course, two LAD arteries running parallel to each other were detected. The artery located on the normal LAD artery region was larger than the other one (courses the apex). The other LAD artery was on the left side of the normal LAD artery and was shorter. Circumflex (Cx) artery was absent. Coronary angiography revealed 70% stenosis of the proximal part of the short LAD; 80% and 50% stenosis on the proximal and mid-segmental part of the long LAD artery, respectively. It resembles to type 2 according to the dual LAD artery anomaly classification (Fig. 1). Right coronary artery (RCA) was superdominant. The Cx artery territory was supplied by the RCA (Fig. 2). An aortography was done in order to detect whether there is a Cx artery or not. The aortography did not show any Cx artery originating from a different ostium.

### Discussion

Dual LAD artery or absence of circumflex anomalies is usually asymptomatic and detected incidentally. Although dual LAD artery anomaly is relatively often, absence of Cx artery is very rare. In this



**Figure 1.** A dual LAD, consisted of a long LAD (black arrow) and a short LAD (white arrow), was demonstrated on the left coronary angiography: A. right 28° and cranial 38° view, B. left 31° and cranial 12° view  
LAD- left anterior descending artery



**Figure 2.** A hyperdominant RCA was also supplying the Cx artery (arrow) on right coronary angiography (left 34°cranial 21° view)  
Cx- circumflex artery, RCA- right coronary artery

case, we detected a combination of type 2 dual LAD artery and absence of Cx artery with the super dominant RCA artery (Fig. 2). The frequency of dual LAD artery is approximately 0.13-1% (4, 5). Spindola-Franco classified dual LAD artery into 4 types (5).

Although absent Cx artery is a much rare anomaly than dual LAD artery, because of co-existing pathologies, absent Cx artery can be symptomatic. Superdominant RCA (Fig. 2) must exist in case of absent Cx. If superdominant RCA does not exist, it must be evaluated for coronary artery hyperplasia syndrome (associated with hypoperfusion and dilate cardiomyopathy) and coronary origin anomaly (Bland-White-Garland syndrome) (3).

According to this classification our case was type 2 dual LAD. Only two cases, in which LAD and Cx artery anomalies were together have been reported (6, 7). Dual LAD artery and Cx artery originating from a different ostium (RCA) was detected in one report (6). In the other report, LAD artery was originating from right coronary cusp and Cx artery was absent (7).

## Conclusion

As far as we know, our case with a combination of the two coronary artery anomalies is the first report in the literature, in which type 2 dual LAD artery and absent Cx has been described in the same patient.

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# Successful treatment of pulmonary arteriovenous malformation and infantile hepatic hemangioendothelioma with alpha-interferon

*İnfanıl hepatik hemanjiyoendotelyoma ve pulmoner arteriyovenöz malformasyonun alfa-interferon ile başarılı biçimde tedavisi*

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## Introduction

Pulmonary arteriovenous malformation (PAVM) is a rare cause of cyanosis in early infancy. PAVMs are direct communications between

the smaller pulmonary arteries and veins (1, 2). Pulmonary arteriovenous malformation in its acquired form usually occurs in juvenile cirrhosis, but has also been reported in patients with trauma, pulmonary schistosomiasis, mitral stenosis, actinomycosis, Fanconi syndrome, and metastatic thyroid carcinoma (3). However, as far as we know infantile hepatic hemangioendothelioma (IHHE) which consists of clinical triad including hepatomegaly, congestive heart failure, and cutaneous hemangiomas with coexistent PAVM has not been reported.

Here, we describe this unusual presentation.

## Case Report

A 10-week old boy was admitted to our hospital from a community hospital for abdominal distension, hepatomegaly and respiratory distress. He developed neonatal jaundice requiring phototherapy on the 3<sup>rd</sup> day of life. Neonatal screening for congenital hypothyroidism demonstrated elevated thyrotropin (TSH) level. He was treated with L-thyroxine.

At the time of referral, the patient was tachypneic, tachycardic and cyanotic with prominent hepatomegaly. The patient was hypoxemic while breathing room air (percent arterial oxygen saturation 70); with oxygen enriched air the percent arterial oxygen saturation raised to 80%. On physical examination he had a heart murmur and congestive heart failure was obtained on echocardiogram. He was started on furosemide and digoxin with rapid stabilization of his cardiac status.

On laboratory evaluation, alpha-fetoprotein (AFP) level was markedly elevated to 2901 ng/ml. The results of blood counts, coagulation studies and liver function tests were all normal. Excretions of catecholamines or metabolites in the urine were within the normal range. Thyroid function tests showed an elevated TSH level of 27.7 mIU/ml. An abdominal sonogram of the patient showed hepatomegaly caused by multiple hypoechoic-isoechoic nodules in both lobes of the liver. Abdominal magnetic resonance imaging (MRI) studies of the hepatic nodules showed decreased signal intensity on T1 images and high signal intensity on T2. There were lesions, up to 3.0 cm in diameter (Fig. 1).

A diagnosis of IHHE was based on the signs of congestive heart failure and hypothyroidism, and the ultrasonography (US) and MRI findings. Then, we started oral prednisolone (2 mg/kg/day) therapy. The patient's hypoxemic and cyanotic status did not change in spite of the therapy. Contrast echocardiography study suggested the presence of intrapulmonary arteriovenous malformation. A contrast echocardiogram showed rapid return of contrast into the left atrium less than 5 seconds after it is seen in the right atrium (Fig. 2) and a diagnosis of diffuse type PAVM was made. We started alpha-interferon 3 million units/m<sup>2</sup> per day over 18 days followed by 3 million units/m<sup>2</sup> 3 times per week on the fifth day of the prednisolone therapy. On the 7<sup>th</sup> day of this combined prednisolone and alpha-interferon therapy his hypoxemic status became normal and the passage of contrast into the left atrium disappeared 1 month after alpha-interferon therapy (Fig. 3). In addition, regression of the hepatic nodules was shown by ultrasound examination approximately 3 months later.

## Discussion

The diagnosis of PAVM should be considered in infants with severe cyanosis without a structural cardiac lesion or pulmonary hypertension, after excluding other causes of cyanosis, such as parenchymal lung disease and the rare methemoglobinemia (1, 4). The gold standard in diagnosis is a pulmonary angiogram showing abnormal peripheral vascular formations that may be either localized or diffuse. Multi-slice computed tomography and contrast echocardiography are also useful. Due to the rapid transit of blood flow through the pulmonary veins into