

## 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.

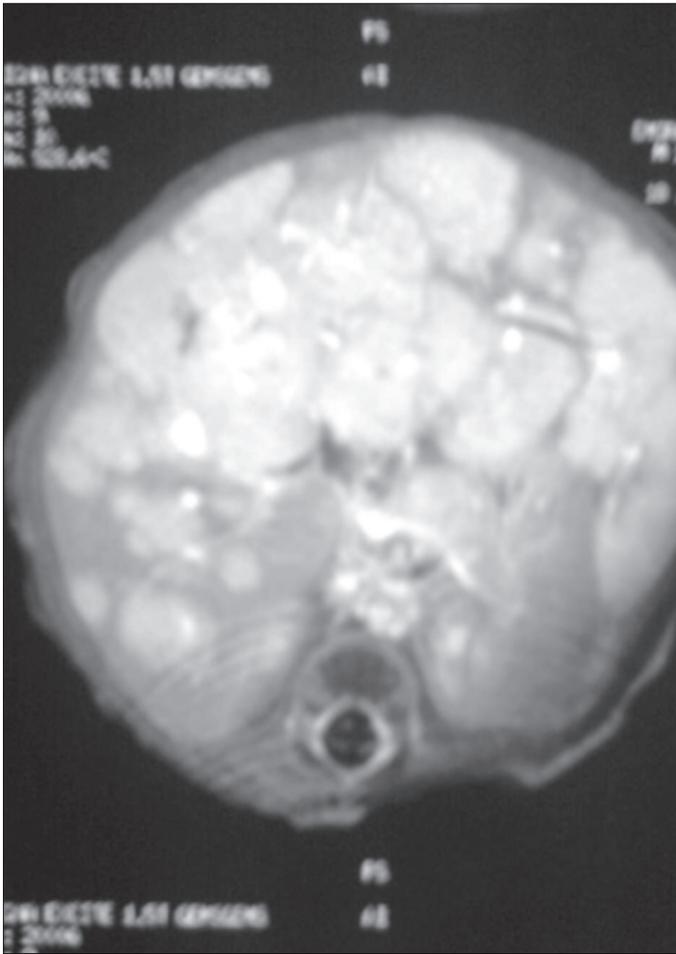
At the time of referral, the patient was tachypneic, tachycardic and cyanotic with prominent hepatomegaly. The patient was hypoxemic while breathing room air (percent arterial oxygen saturation 70); with oxygen enriched air the percent arterial oxygen saturation raised to 80%. On physical examination 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.

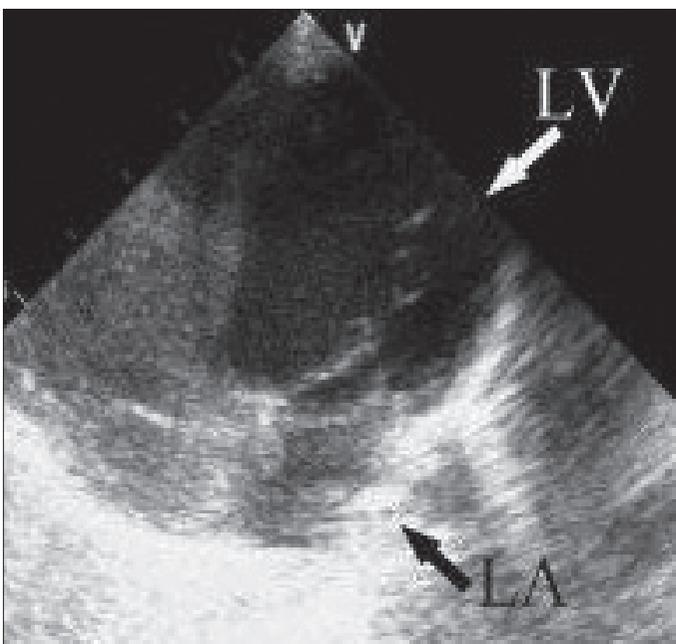
## Discussion

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

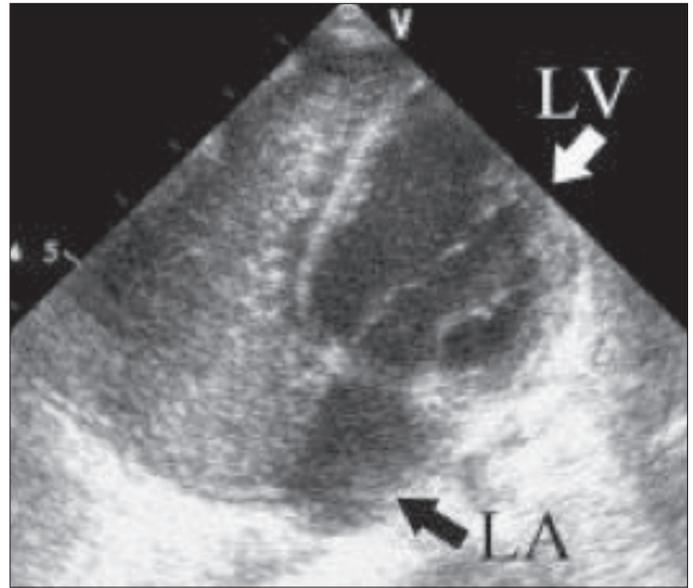


**Figure 1. T1-weighted MRI shows multiple lesions up to 3 cm diameter in the liver**

MRI - magnetic resonance imaging



**Figure 2. Contrast echocardiogram shows rapid return of contrast into the left atrium less than 5 seconds after it is seen in the right atrium**



**Figure 3. Contrast echocardiogram shows no rapid contrast passing through into the left atrium after treatment**

the capillaries, the left atrium is seen to be filled early with contrast echoes (within 2-5 seconds) following contrast passage through the right atrium, as we found in our case (5-7).

Treatment of localized PAVMs includes surgical lobectomy or transcatheter embolization using coils, but there is no definitive treatment for the diffuse type. Interferon therapy has been used in patients PAVMs with juvenile cirrhosis (8-10). However, until the present time, its efficacy for the treatment of PAVM has not been clearly demonstrated. In our case, oral prednisolone was initially used because of the extensive and multiple lesions of IHHE. After the diagnosis of PAVM, we added interferon alpha to the prednisolone therapy based on the assumption that interferon alpha is beneficial for both of the illness.

### Conclusion

Pulmonary arteriovenous malformation should be kept in mind in cyanotic infants with IHHE. Contrast echocardiography may be a useful non-invasive technique for detecting PAVM and the interferon alpha may be first treatment option for patients with IHHE and PAVM.

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## Echocardiographic diagnosis of atrioventricular septal defect without primum atrial septal defect: a relatively "common" congenital heart defect in Down's syndrome

*Primum atriyal septal defekt olmaksızın, atriyoventriküler septal defektin ekokardiyografik tanısı: Down sendromunda görece "yaygın" konjenital kalp defekti*

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### Introduction

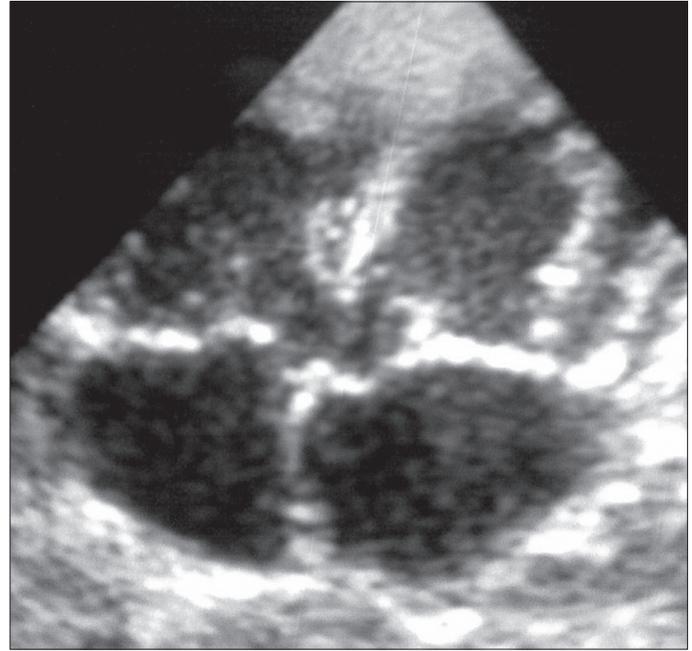
Atrioventricular septal defects (AVSD) are congenital heart defects characterized by a common atrioventricular (AV) junction guarded by an abnormal valve with variable presence of primum atrial septal defects (ASD) and inlet ventricular septal defects (VSD) (1). A less common form of AVSD occurs when the common leaflet is fused to either the atrial septum or to the crest of the ventricular septum, creating an AVSD with no interatrial or interventricular communications respectively. There have been case reports of AVSD with intact septa, where the diagnosis was made at necropsy (2). In 2005, we validated quantitative echocardiographic measurements that helped in diagnosing of AVSD with no primum ASD: the measurements of the left ventricle inlet to outlet distance ratio and the percentage of the left AV valve guarded by the mural leaflet (3). In this report we describe four more cases of AVSD without primum ASD diagnosed using the above echocardiographic measurements.

### Case Report

Four patients with AVSD and intact atrial septum were identified out of 50 patients with Down's syndrome and AVSD (3.7%).

All four patients were males. Their ages were 2, 3, 12 and 36 months. They all presented with shortness of breathing and failure to thrive. In all patient's physical examination revealed signs of congestive heart failure which was mild in the 36-month-old patient. Echocardiographic examination was done and revealed the following:

1. A large inlet VSD
2. No primum ASD (Fig. 1)
3. A common atrioventricular (AV) junction with a common AV valve (Fig. 2)
4. The percentage of the left AV valve guarded by the mural leaflet was 30-40 (Fig. 3A and B)



**Figure 1. A four -chamber echocardiographic view showing a large inlet VSD with no primum ASD. Abnormal AV valve noted**

ASD - atrial septal defect, AV - atrioventricular, VSD - ventricular septal defect



**Figure 2. En-face view of the same patient in Figure 1 showing a common AV junction with a small mural leaflet occupying 32% of the left AV valve orifice**

AV - atrioventricular