

hypertension, trombophilia, anemia, diabetes mellitus, smoking and preeclampsia.

### Conclusion

Acute MI during pregnancy has high maternal and fetal mortality rates and in most cases vasospasm, hypercoagulable state and additional exogenous factors (e.g. progestogens and smoking) may be the underlying mechanism.

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## Hepatopulmonary syndrome associated with Budd-Chiari syndrome

### *Budd-Chiari sendromu ile birliktelik gösteren hepatopulmoner sendrom*

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### Introduction

Budd-Chiari Syndrome (BCS) is a rare cause of portal hypertension. Occlusion of inferior vena cava (IVC) or hepatic vein (HV) causes BCS and leads to centrilobular congestion and necrosis of liver. We present a cyanotic patient with BCS associating with hepatopulmonary syndrome (HPS).

### Case Report

A 15-year-old girl admitted with dyspnea and cyanosis. She was referred to our clinic with the prediagnosis of congenital heart disease. On examination, there was cyanosis of the skin and mucosa; clubbing of the fingers and toes (Fig.1) and a grade 2/6 systolic murmur. Electrocardiography showed right axis deviation. Chest X-ray and thoracic computed tomography findings are presented in Figure 2. The pulmonary veins in the lower parts of the lungs were prominent and enlarged.

Her laboratory results revealed; hemoglobin, 17.9 g/dl; platelets, 120000/mm<sup>3</sup>, white blood cell count, 5000/mm<sup>3</sup>. Blood glucose level was 73 mg/dl, alanine aminotransferase, 29 U/L; aspartate aminotransfer-

ase, 38 U/L; gamma-glutamyl transferase, 52 U/L; alkaline phosphatase, 490 U/L; total protein, 7.6 mg/dl; albumin, 3.9 mg/dl; total bilirubin, 3.32 mg/dl; direct bilirubin, 1.02 mg/dl. Arterial blood gas analysis revealed;



**Figure 1. Cyanosis and clubbing of the fingers are shown in the figure**

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doi:10.5152/akd.2010.073

pH, 7.403; PCO<sub>2</sub>, 30.9 mmHg; PO<sub>2</sub>, 31.1 mmHg; SO<sub>2</sub>, 54.9%; HCO<sub>3</sub>, 18.9 mmol/L; MetHb, 0.9 %. Hepatitis A, B and C, cytomegalovirus and toxoplasma antibodies were negative. Paroxysmal nocturnal hemoglobinuria panel,  $\alpha$ 1-antitrypsin, blood copper, ceruloplasmin, sweat test, protein C and S, antithrombin III were in normal limits. Factor V Leiden mutation, prothrombin 20210A mutation were negative.

After echocardiography revealed normal findings, contrast-enhanced echocardiography was performed. Contrast medium, seen as micro bubbles, after appeared in right chambers, appeared in left heart in a period of 4-6 heart beats.

Oxygen saturation by digital pulse oximetry was 72-74%. With O<sub>2</sub> (2lt/min, nasal cannula), it was 78%. It was 65-69 % in upright position and 50-55 % with exertion.

Pulmonary angiography showed dilated capillaries (Fig. 3). On abdominal ultrasound and CT (Fig. 4a), multiple large nodules in liver and splenomegaly were seen (vertical length: 150 mm). Esophagogastroduodenoscopy revealed esophageal varicose veins. Doppler ultrasound showed normal course of IVC. Liver biopsy revealed prominent dilatation of sinusoids around central veins (Fig. 5) which was compatible with venous outflow obstruction. Vena cavography and hepatic venography showed that there was not a web in IVC and the course of right HV and its branches were abnormal (Fig. 4b). Partial liver transplantation was planned for patient.

## Discussion

For patients with cyanosis, intracardiac and intrapulmonary shunts should be considered. Contrast-enhanced echocardiography is useful for discrimination (1).

Differential diagnosis of intrapulmonary shunts includes two conditions: pulmonary arteriovenous malformations (PAVM) and HPS (2). PAVM are characterized by abnormal communications between the pulmonary arteries and veins (3). Approximately two thirds of PAVM occur in hereditary hemorrhagic teleangiectasia (2).

The triad of 'HPS' are liver disease, hypoxemia with a PaO<sub>2</sub><70 mmHg while breathing room air and evidence of intrapulmonary vascular dilatations (4). Patients with HPS can present with either hepatic (80%) or pulmonary symptoms (20%) (5). Most common pulmonary symptom is dyspnea, which may accompany platypnea, and/or orthodeoxia. Platypnea and orthodeoxia are not pathognomonic for HPS. But, association with liver dysfunction strongly suggests HPS.

Hepatopulmonary syndrome (HPS) is associated with many types of liver diseases. Association with BCS is rare (6). Budd-Chiari Syndrome (BCS) which is due to hepatic outflow obstruction, occur in a variety of conditions, particularly prothrombotic states. Occlusion of a single HV is usually silent (7). Overt BCS generally requires the occlusion of at least two HV. Enlargement of the caudate lobe is common because blood is shunted through it directly into IVC. Large nodules in liver have been described in literature as benign regenerative nodules associated with BCS (8). In our patient, symptoms of HPS were dominant. BCS showed a severe and chronic course because of insidious progress of HPS. Caudate lobe hypertrophy, large nodules in the liver, hepatic venographic findings and liver biopsy were compatible with BCS.

In series of Gentil-Kocher et al. (9), all children with BCS had hepatomegaly and 3 children had acute refractory ascites. Liver function tests were normal in most of them. Dilawari et al. (10) suggested that children usually do not have acute BCS, and chronic BCS in children and adolescents is similar to BCS in adults.



Figure 2. a) Chest radiogram b) Chest tomography of the basal segment

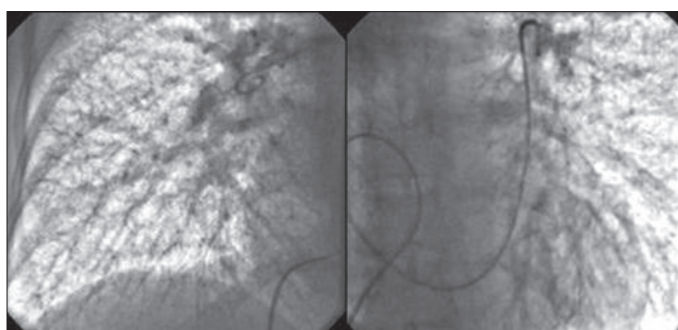


Figure 3. Pulmonary angiogram of the patient shows the extensive diffusely dilated spider-like capillaries. This appearance, together with the poor response to 100% inspired oxygen, fits to the type 1 diffuse HPS  
HPS - hepatopulmonary syndrome

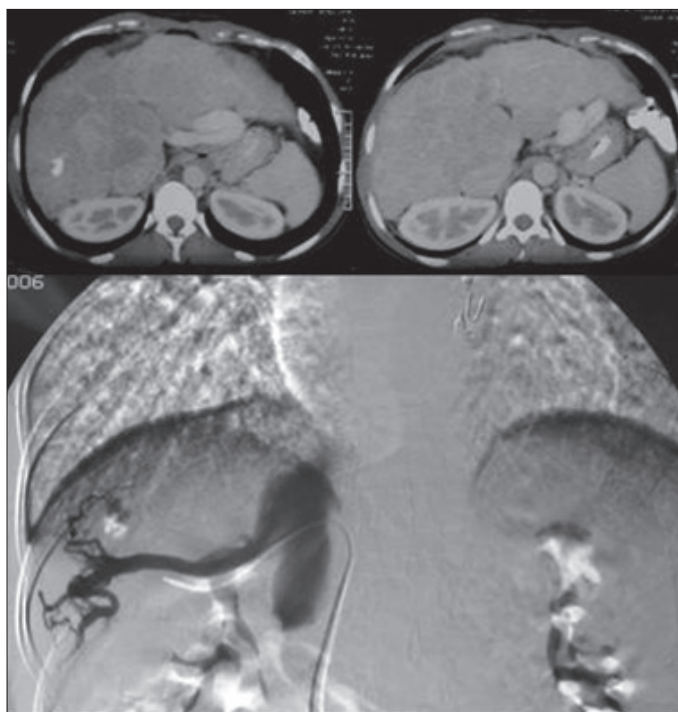
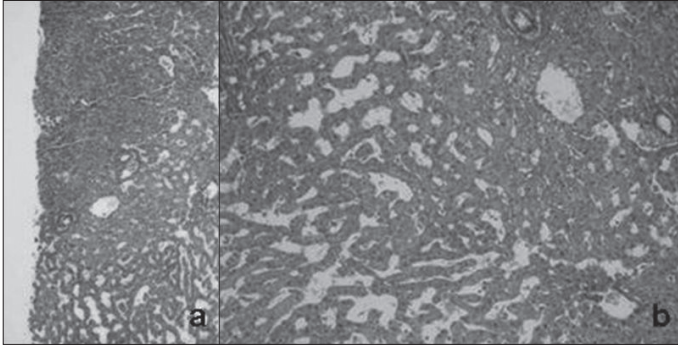


Figure 4. a) Abdominal tomography shows the lobulation of the liver b) Hepatic venography: the course of the right hepatic vein is abnormal, and the middle and the left hepatic veins are not seen



**Figure 5. Dilatations of sinusoids around the central veins (Trichrome stain, a. x40 and b. x100)**

### Conclusion

This is a rare case of BCS that showed clinical features of HPS before clinical findings of liver dysfunction. Because of her deep cyanosis due to HPS, differential diagnosis with cyanotic congenital heart diseases, Eisenmenger syndrome and pulmonary hypertension was required.

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## Late onset LIMA first branch steal syndrome after coronary artery bypass surgery

*Koroner arter baypas cerrahisi sonrası geç ortaya çıkan LIMA ilk dal çalma sendromu*

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### Introduction

Left internal mammary artery (LIMA) use for left anterior descending artery (LAD) in coronary artery bypass surgery (CABG) has been accepted as the first choice of graft due to its long term high patency rate (1). Histologically, the artery has a strong elastic membrane, which helps the vessel resist the atherosclerotic process (2). On the other hand, postoperative angina is not always related to arteriosclerotic coronary stenosis. Occasionally, an untied LIMA branch may redirect the blood flow toward the thoracic wall and can cause angina pectoris (3).

In this report, we present a patient who presented with late onset of angina pectoris related to the untied first branch of LIMA after CABG operation. The patient was operated using a minimally invasive technique.

### Case report

A 71-year-old-male was admitted to our clinic with new onset of effort related angina pectoris. The patient had known hypertension, hyperlipidemia and coronary artery disease with a two-vessel coronary artery bypass surgery done (LIMA to left anterior descending artery (LAD), saphenous vein graft (SVG) to the second marginal branch (OM2)) 2 years ago. Reversible ST segment elevation in the anterior precordial leads was detected on exercise treadmill test electrocardiogram (ECG). The ejection fraction was 55% with no wall motion abnormalities noted. On control coronary angiography, new coronary artery lesions were not observed and LIMA-LAD, SVG-OM2 grafts were seen to be patent.

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doi:10.5152/akd.2010.074