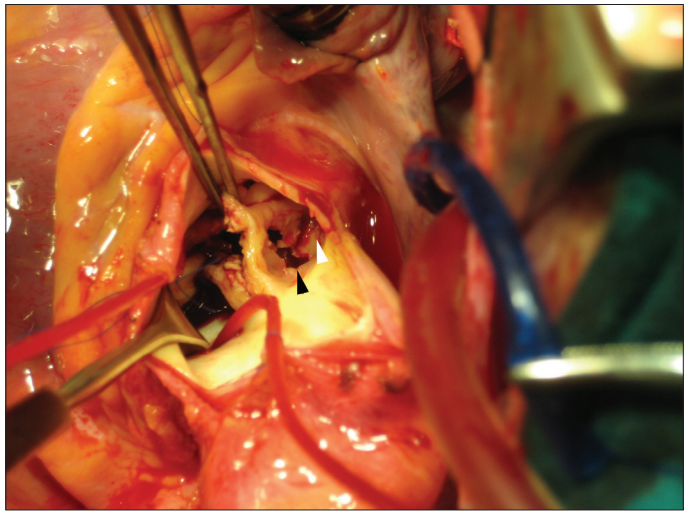

Figure 2. The tract of the fistula and the rupture on the posterior leaflet of the bicuspid aorta: intraoperative view

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Ventricular septal defect with bidirectional shunting in a patient with congenitally corrected transposition

Konjenital düzeltilmiş transpozisyonlu bir hastada bidireksiyonel şantlı ventriküler septal defekt

Congenitally corrected transposition of the great arteries (CCTGA) is a rare cardiac malformation characterized by the combination of discordant atrioventricular and ventriculoarterial connections. Most of the cases with CCTGA are diagnosed in childhood because of concomitant cardiac malformation. Relevant concomitant cardiac defects such as ventricular septal defect (VSD), atrial septal defect, tricuspid regurgitation and pulmonary stenosis were reported previously. We report an asymptomatic patient with CCTGA and coexisting VSD with bidirectional shunting.

A 22-year-old asymptomatic male in the army was seen in our department during his periodical examination. He had a grade 3/6 mesocardiac systolic murmur on cardiac auscultation. Electrocardiogram (ECG) showed normal sinus rhythm with right bundle branch block. Transthoracic echocardiography showed CCTGA with VSD with left to right shunt (Fig.1, Video 1. See corresponding video/movie images at www.anakarder.com) and moderate tricuspid and aortic regurgitation in apical four-chamber view. The pulmonary valve was moderately stenotic with a peak pressure gradient of 49 mm Hg. For identifying the direction of shunt flow in VSD contrast echocardiographic examination with agitated saline was carried out. Contrast echocardiography demonstrated positive contrast effect in the left ventricular in diastole confirming a right-to-left shunt at the ventricular septum (Video 2. See corresponding video/movie images at www.anakarder.com). According to our knowledge, our case is the first reported CCTGA with VSD with bidirectional shunting in an asymptomatic patient.

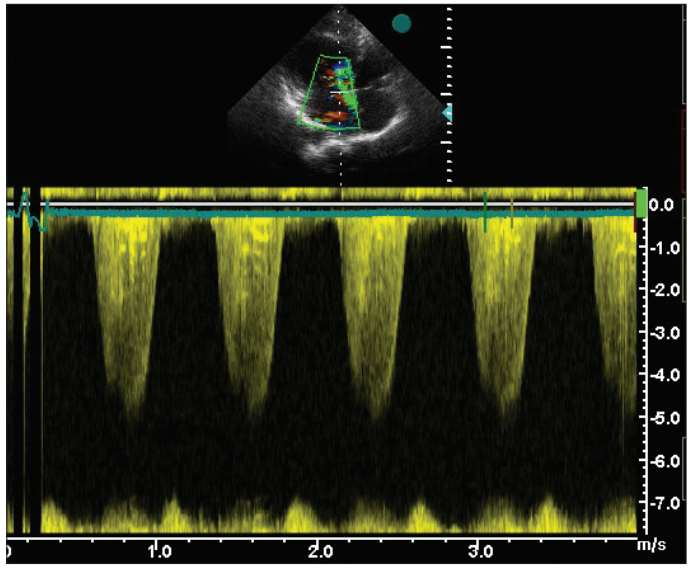

Figure 1. Transthoracic and Doppler echocardiography views of congenitally corrected transposition of the great arteries with ventricular septal defect with left to right shunt

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Huge main pulmonary arterial thrombus in a child with increased lipoprotein (a) level

Lipoprotein (a) yüksekliği olan bir çocukta pulmoner arteriyel dev trombüs

Pulmonary arterial thrombosis is an extremely rare clinical condition both in children and in adults. Lipoprotein (a) [Lp (a)] is an atherogenic

lipoprotein particle which displays adjunctive thrombotic properties by inhibition of the fibrinolytic pathway.

A 9-year-old boy with no previous history of cardiac or pulmonary disease was referred to our hospital for investigation of dyspnea and tachypnea that had started one month ago. In the echocardiogram and computed tomography (CT)-angiogram, a large thrombus was seen in the main pulmonary artery bifurcation which almost completely obstructed the right pulmonary artery and partially the left one with tricuspid regurgitation and dilation of the right chambers of the heart (Fig. 1, 2).

Thrombophilia screening was normal except high Lp (a) concentration (1.33 g/L, normal: 0.01-0.30 g/L). We performed family screening for Lp (a) and found a high Lp(a) level (0.9 g/L) in his father. While the patient was on the heparin therapy; a sudden increase in his tachypnea, dyspnea and anxiety occurred and he complained of pleuritic chest pain. Since pulmonary perfusion scintigraphy with Tc^{99} macroaggregated albumin revealed bilaterally perfusion defects in the lungs, the patient was immediately referred for thrombectomy and the thrombus was removed totally (Fig. 3). As a result, he was discharged from the hospital under warfarin and low-dose aspirin therapy.

Elevated Lp (a) is a very rare cause of venous and arterial thromboembolism and should be checked in such cases both to determine the etiology of the thrombus and for detecting the other family members with increased Lp (a) level and thromboembolism risk.

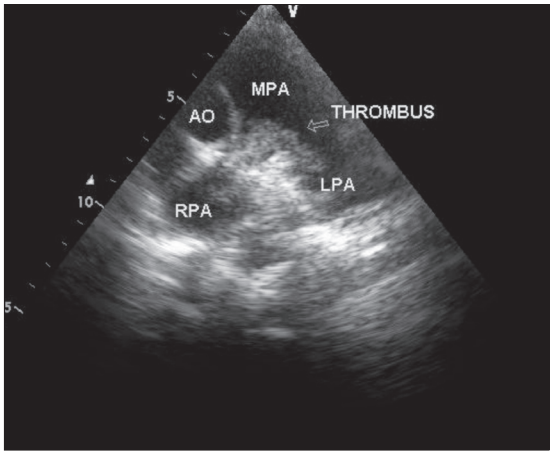

Figure 1. Transthoracic parasternal short-axis echocardiography view of a large thrombus in the main pulmonary artery bifurcation
AO-aorta, LPA-left pulmonary artery, MPA - main pulmonary artery, RPA- right pulmonary artery

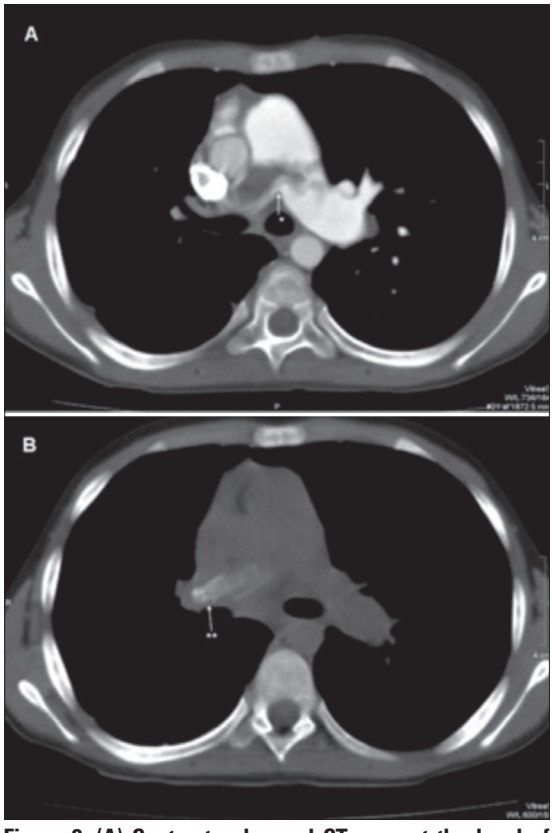

Figure 2. (A) Contrast enhanced CT scan at the level of bifurcation of pulmonary arteries reveals low - density thrombus located mainly in the right pulmonary artery (*) (B) Unenhanced CT scan shows calcified thrombus (**)
CT - computerized tomography

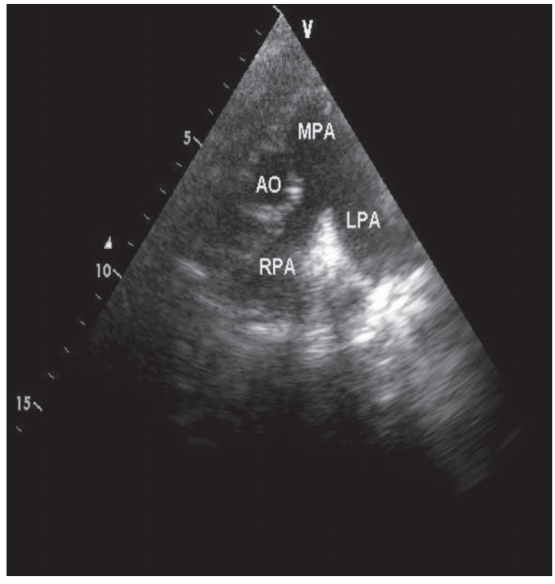

Figure 3. Transthoracic parasternal short - axis echocardiographic view of the main pulmonary artery after thrombectomy
AO-aorta, LPA - left pulmonary artery, MPA - main pulmonary artery, RPA - right pulmonary artery

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