

## A patient with Turner's syndrome associated with unexplained left ventricular hypertrophy, severe left ventricular systolic dysfunction, atrial septal defect and pericardial effusion

*Açıklanamayan sol ventrikül hipertrofisi, ciddi sol ventrikül sistolik disfonksiyonu, atriyal septal defekt ve perikardiyal efüzyon birlikteliği olan Turner sendromlu bir hasta*

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A 40-year-old female with Turner's syndrome presented with a 3 months history of progressive dyspnea and peripheral edema. Physical examination revealed signs of biventricular failure. Blood pressure was 110/70 mmHg. The patient had been seen by physicians previously and there had been no signs of hypertension. Electrocardiogram showed left and right atrial dilatation, left ventricular hypertrophy and ST depressions in the infero-lateral derivations. Transthoracic echocardiography images revealed left ventricular hypertrophy (interventricular septum: 15 mm, posterior wall: 22 mm), severe left ventricular systolic dysfunction (ejection fraction: 10%), dilated left atrium (50 mm), moderate pericardial effusion (Fig. 1), secundum atrial

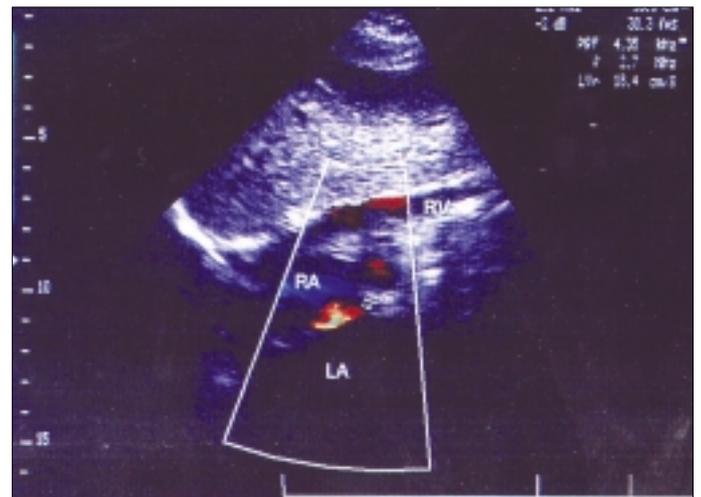
septal defect (Fig. 2), moderate mitral and tricuspid regurgitations, pulmonary hypertension (pulmonary artery pressure: 50 mmHg), left ventricular diastolic dysfunction (mitral A wave>E wave) and dilations of the right heart chambers. A medical therapy including beta-blocker, diuretic, angiotensin converting enzyme inhibitor and digitalis was commenced.

There is high prevalence of congenital heart defects in patients with Turner's syndrome. Aortic malformations, including bicuspid aortic valve, coarctation of the aorta and aortic dilation are the most common. Hypertension, which may cause left ventricular hypertrophy is frequent in Turner's syndrome (1). However there was no hypertension in our



**Figure 1. Transthoracic echocardiography, parasternal long-axis view showing hypertrophied left ventricle, pericardial effusion and dilated left atrium. Arrow indicates pericardial effusion**

AO- aorta, LA- left atrium, LV- left ventricle, RV- right ventricle



**Figure 2. Transthoracic echocardiography, subcostal view. Interatrial septal defect is shown. Arrow indicates left to right atrial shunt**

LA - left atrium, RA- right atrium, RV- right ventricle

patient. The patient had been seen by physicians previously and there had also been no hypertension as a cause of left ventricular hypertension. Although atrial septal defect is frequent in Noonan syndrome, it is extremely rare in Turner's syndrome (2). Pericardial effusion has never been reported in Turner's syndrome.

## References

1. Lin AE, Lippe B, Rosenfeld RG. Further delineation of aortic dilation, dissection, and rupture in patients with Turner's syndrome. *Pediatrics* 1998; 102: e12.
2. Pyeritz RE. Genetics and cardiovascular disease. In: Braunwald E, Zipes DP and Libby P, editors: *Heart Disease: a Textbook of Cardiovascular Medicine*. 6th ed. Philadelphia: WB Saunders; 2001. p.1977-2010.