

Cor triatriatum sinister — a rare cause of right heart failure

Vlatka Rešković Lukšić*, Margarita Brida, Blanka Glavaš Konja, Joško Bulum, Alexander Ernst, Darko Anić, Jadranka Šeparović Hanževački

University of Zagreb School of Medicine, University Hospital Centre Zagreb, Zagreb, Croatia

A 40-year-old male was admitted to our Clinic for evaluation of pulmonary arterial hypertension. Transthoracic (TTE) and transesophageal (TEE) two and three-dimensional echocardiography revealed obstructive membrane in the left atrium with small fenestration of only few millimeters, pulmonary artery pressure (PAP) of 60 mmHg and moderately dilated and hypertrophied right ventricle (RV). Late presentation led us to look for concomitant anomalies, so a partial anomalous left pulmonary venous connection was found. The diagnosis was confirmed by computed tomography (MSCT) — upper left pulmonary vein (LUPV) was drained into left brachiocephalic vein via vertical vein, and no lung anomalies were found. On coronary angiography, separate origins of the left anterior descending and circumflex artery were shown. The patient was referred to a cardiac surgeon, but he refused an operative correction at the time. Three years later, he was admitted because of dyspnea on exertion and clinical signs of right sided heart failure. On TTE, impairment of RV function was found, with severe RV dilatation and PAP of 90 mmHg. Cardiac magnetic resonance revealed no sig-

nificant amount of RV fibrosis, and by right heart catheterization, pulmonary vascular resistance index (PVRI) of 1.45 Wood was calculated. Left-to-right shunt was estimated by radionuclide angiography to 26-30%. After medical stabilization, surgical resection of intraatrial membrane and LUPV reconnection into the left atrium appendage was performed. One month after the operation, the patient was asymptomatic and positive remodeling of RV, normal flow in the left atrium and PAP of 23 mmHg were found.

Cor triatriatum sinister is a very rare condition, especially in combination with anomalous pulmonary venous connection, so it is essential to search for concomitant anomalies. In the patient with signs of RV failure, multimodality approach was used to estimate the RV function. The calculation of PVRI was very important in making a decision whether this patient was operable. Some congenital causes of RV heart failure, such as cor triatriatum with anomalous pulmonary venous connection are important to be recognized in adults since they are surgically correctable due to low PVR. Apart from the diagnosis, multidisciplinary approach is also necessary in the assessment of RV function and PVR, key determinants in successful operation.

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*Address for correspondence: Klinički bolnički centar Zagreb, Kišpatičeva 12, HR-10000 Zagreb, Croatia.

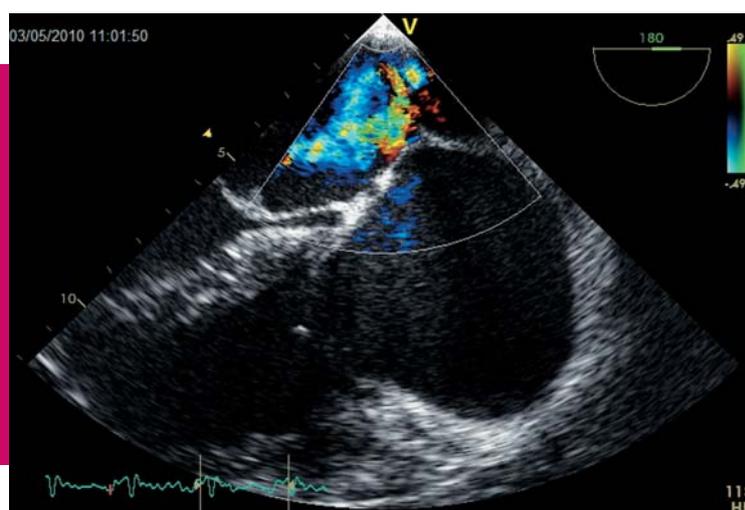
Phone: +385-91-5612-526

E-mail: vlatka.reskovic@gmail.com

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Figure 1. The transesophageal echocardiogram: obstructive membrane in left atrium. Small fenestration with turbulent flow is seen on the top of the picture.



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