Sinus venosus atrial septal defect with anomalous right superior pulmonary vein inflow into the vena cava superior in a middle-aged woman: a case report

Ivona Mustapić[1,2]*, **Zora Sušilović Grabovac**[1,2], **Darija Baković Kramarić**[1,2]

[1]University of Split School of Medicine, Split, Croatia
[2]University Hospital Centre Split, Split, Croatia

KEYWORDS: atrial septal defect, congenital heart disease, echocardiography.

CITATION: Cardiol Croat. 2021;16(5-6):187. | https://doi.org/10.15836/ccar2021.187

***ADDRESS FOR CORRESPONDENCE:** Ivona Mustapić, University Hospital Centre Split, Spinčićeva 1, 21000 Split, Croatia. / Phone: +385-95-227-6970 / E-mail: mateljanka@gmail.com

ORCID: Ivona Mustapić, https://orcid.org/0000-0002-1534-3642 • Zora Sušilović Grabovac, https://orcid.org/0000-0001-9999-7557 Darija Baković Kramarić, https://orcid.org/0000-0001-6751-5242

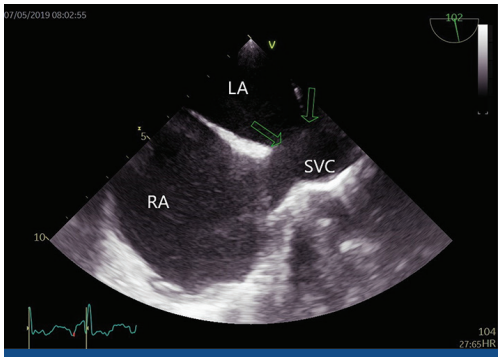

FIGURE 1. Transesophageal echocardiography. Arrow pointing at a superior sinus venosus atrial septal defect.
LA - left atrium; RA - right atrium; SVC - superior vena cava.

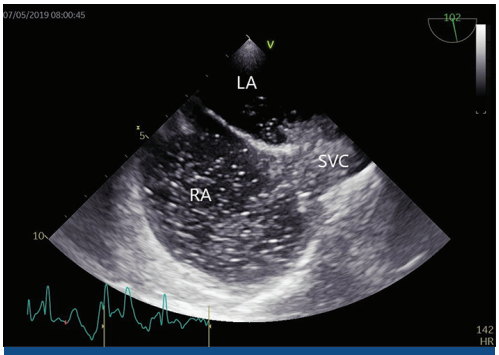

FIGURE 2. Transesophageal echocardiography with contrast study.
LA - left atrium; RA - right atrium; SVC - superior vena cava.

Introduction: Atrial septal defect (ASD) is the second most common adult congenital heart disease, usually asymptomatic until the third decade. Superior sinus venosus defect (SVASD) account for 5% of ASD and it is usually associated with the partial or complete connection of right pulmonary veins to vena cava superior (SVC) or right atrium (RA)[1,2].

Case report: We report a case of a 45-year-old woman with previously known thyroiditis and hyperprolactinemia. She was referred for echocardiographic examination after an accidental finding of mid-systolic murmur during preoperative preparation for ovarian cyst surgery. She worked as a waiter and reported exertional dyspnea. An electrocardiogram revealed sinus rhythm with the right bundle branch block. Transthoracic echocardiography (TTE) demonstrated a normal-sized left heart with preserved systolic function (LVEF 70%), a dilated RA and right ventricle (RV) without signs of pulmonary hypertension. Cardiac magnetic resonance (CMR) showed a dilated RV (end-diastolic diameter 46 mm), dilated RA and dilated pulmonary artery (diameter 33 mm). During the CMR scan there was constantly a high concentration of contrast in the RV which raised suspicion of shunt presence. Transesophageal echocardiography using contrast revealed SVASD (**Figure 1**, **Figure 2**). The patient was referred for computer tomography angiography which demonstrated superior SVASD, 16 mm in width. The right superior pulmonary vein had abnormal inflow into SVC, while the right inferior and both left pulmonary veins had typical anatomical inflow into the left atrium. Cardiac scintigraphy with technetium-99 confirmed the existence of a left-right shunt, Qp: Qs ratio of 1.7: 1. Surgical repair was performed by forming an intraatrial patch using autologous pericardium and dilating plastic of the SVC and RA with a xenopericardial patch. Postoperative recovery went well and control TTE showed less dilated RV with good patch position and no signs of shut over intraatrial septum.

Conclusion: We represented a rare case of congenital heart disease, diagnosed in a middle-aged patient. Having a patient with a dilated right heart and normal-sized left heart without signs of pulmonary hypertension needs to raise suspicion of L-R shunt existence and further investigation should be done.

RECEIVED:
March 28, 2021

ACCEPTED:
April 2, 2021

LITERATURE

1. Baumgartner H, De Backer J, Babu-Narayan SV, Budts W, Chessa M, Diller GP, et al; ESC Scientific Document Group. 2020 ESC Guidelines for the management of adult congenital heart disease. Eur Heart J. 2021 Feb 11;42(6):563-645. https://doi.org/10.1093/eurheartj/ehaa554
2. Martin SS, Shapiro EP, Mukherjee M. Atrial septal defects - clinical manifestations, echo assessment, and intervention. Clin Med Insights Cardiol. 2015 Mar 23;8(Suppl 1):93-8. https://doi.org/10.4137/CMC.S15715