Background: Systemic sclerosis (SSc) is a connective tissue disease characterized by fibrosis and vascular remodelling with poor long-term survival of patients who develop pulmonary arterial hypertension (PAH). Exercise echocardiography might improve the early diagnosis of PAH by unmasking exercise-induced PAH (ex-PAH) but is not feasible in all SSc patients. We sought therefore to investigate whether pulmonary artery enlargement and right ventricle (RV) dysfunction were parameters indicative of ex-PAH.

Methods: 89 SSc patients with normal resting systolic pulmonary artery pressure (sPAP) and without severe pulmonary function abnormalities underwent exercise echocardiography and multislice computed tomography (MSCT). They were divided into two groups according to the presence of ex-PAH, defined by sPAP >45 mmHg. Right ventricular (RV) function was evaluated using 2D strain. The ratio of main pulmonary artery diameter (mPA) over the diameter of ascending aorta (Ao) was determined using MSCT.

Results: As compared with non ex-PAP group, ex-PAH group was older (60 ±10 vs 50 ±13 years, P=0.001), had higher exercise sPAP (54 ±10 vs 34 ±6 mmHg, P<0.001), increased mPA diameter (33.6 ±4.5 vs 30.6 ±3.9 mm, P=0.003) and mPA/Ao ratio (1.1 ±0.1 vs 1.0 ±0.1, P=0.001). Global RV free wall strain was lower in ex-PAH group than in non ex-PAP group (24 ±5 vs 30 ±5%, P<0.001). Exercise sPAP was significantly correlated with mPA/Ao ratio. Sensitivity and specificity of mPA/Ao ratio >1 to identify ex-PAH was 80% and 81%, respectively. Multivariate analysis identified age, RV strain and mPA/Ao ratio as independent parameters indicative of ex-PAH.

Conclusions: In SSc patients with normal resting sPAP, increase in mPA/Ao ratio and decreased RV strain are parameters that can indicate the presence of exercise PAH.

KEYWORDS: pulmonary artery hypertension, pulmonary artery diameter, speckle tracking echocardiography, systemic sclerosis.

Figure 1. Predictive factors of exercise pulmonary artery hypertension.

Prošireni sažetak / Extended abstract

Exercise pulmonary hypertension is associated with increased pulmonary artery dimension in systemic sclerosis

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