

Pulmonary Hypertension in Patients with Juvenile Lupus Erythematosus

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Abstract: Introduction:

Juvenile systemic lupus erythematosus (jSLE) is a chronic multisystemic autoimmune disease characterized with variable clinical course. Vital organ involvement is the most important morbidity and mortality factor of the disease. Pulmonary hypertension (PH) is defined as a mean pulmonary artery pressure (PAP) ≥ 25 mmHg at rest or a right ventricular systolic pressure (RVSP) > 40 mmHg. Non-specific clinical features and insignificant findings are the main reasons of the delayed diagnosis in patients with PH. Timely echocardiographic (ECO) examination enables early diagnosis. Pulmonary hypertension has been reported to be associated with poor prognosis in several studies among adults with SLE. The prevalence of PH in adult SLE patients is estimated to be 1.8% to 14%. Studies on pulmonary hypertension among patients with jSLE are spare.

Objectives:

The aim of this study is to explore the right ventricle functions and to determinate the frequency of pulmonary hypertension in patients with jSLE, using non-invasive methods (Pulsed wave and tissue Doppler ECO).

Methods:

Patients with diagnosis of jSLE followed up at our department were included in the study. Pulse wave and tissue Doppler ECO was performed to all included patients and to healthy controls. A complete medical history and laboratory results were taken from patients' medical records. Physical examination was performed for both studied patients and healthy controls, in order to exclude the coincidental disease. Measurements obtained by ECO include: peak velocity of the tricuspid insufficiency (TRVmax), end diastolic velocity of pulmonary insufficiency (PIV), tricuspid annular plane systolic excursion (TAPSE), right ventricle diastolic function measurement (Lat E, A, E' wave, E/E' ratio). Bernoulli equation ($4 \times \text{TRVmax}^2$) was used to calculate the estimated PAP. Student's t-test was used for data with normal distribution. Mann-Whitney test was used to assess abnormally distributed continuous data. Fisher's exact test was used to compare categorical data. Pearson's correlation was used to investigate Doppler echocardiographic measurements.

Results:

A total of 38 jSLE patients and 40 healthy controls were included in the study. Mean age of patients was 16.0 ± 2.59 years, mean age at diagnosis was $10,63 \pm 3,51$ years and mean disease duration was 57.02 ± 33.6 months. Mean age of control group was 15 ± 3.49 years. TRV max and PIV were significantly higher in jSLE patients comparing to healthy controls with $p < 0.05$ and $p < 0.001$, respectively.

Mean TRVmax was $2,340 \pm 0,277$ m/sec in jSLE and $2,044 \pm 0,411$ m/sec in healthy controls. Mean PIV was $1,469 \pm 0,295$ and $1,214 \pm 0,128$ m/sec in jSLE and healthy controls, respectively. Tissue Doppler ECO measurements of right cardiac diastolic functions (Lat E, E' wave and E/E' ratio) were significantly different in jSLE patients, comparing to healthy controls. Lat E wave (cm/sec) was $13,441 \pm 1,463$ and $16,767 \pm 2,605$, Lat E' wave was

6,772±1,297 and 5,875±0,720 for jSLE patients and healthy controls, respectively. Lat E/E' ratio was 5,227±1,192 in jSLE patients and 5,841±0,677 in healthy controls.

Conclusion:

This study confirms that pulmonary hypertension is uncommon among patients with jSLE. However, patients with jSLE have a compromised right cardiac contractile functions and higher pulmonary artery pressure comparing to healthy controls. These results point out the importance of echocardiographic examination in patients with jSLE, regarding right cardiac functions and pulmonary hypertension.

Key words: juvenile systemic lupus erythematosus, pulmonary hypertension