中文題目：結締組織疾病相關之肺動脈高壓患者長期存活分析

英文題目：Long-term survival of patients with connective tissue disease-associated pulmonary arterial hypertension: A single-center cohort

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Background: Pulmonary arterial hypertension (PAH) is a rare but severe complication of connective tissue disease (CTD). CTD-associated PAH (CTD-PAH) is the most common type among East Asians.

Method: We analyzed 41 patients with CTD-PAH who were enrolled over the past 15 years. Systemic lupus erythematosus (SLE) was the most common etiology, followed by systemic sclerosis (SSc) and mixed CTD (MCTD). All patients completed follow-up visits every 3–6 months to record clinical symptoms, hematologic and biochemical tests, 6-minute walk distance (6MWD), and echocardiographic examination.

Results: Among the 39 (95%) patients who received PAH-specific therapy, 24 (59%) received monotherapy and 15 (37%) received combination therapy. The following parameters were observed during the follow-up visits: improved World Health Organization functional class, 6MWD, uric acid levels, estimated systolic pulmonary artery pressure (SPAP), tricuspid annular plane systolic excursion, and ratio of tricuspid regurgitation velocity and velocity time integral at the right ventricular outflow tract. Increased C-reactive protein (CRP) levels were frequently observed. Baseline CRP level, SPAP, main pulmonary artery diameter, and pulmonary vascular resistance were higher in the non-survival than in the survival group. The 1-, 2-, 3-, and 5-year survival rates were 90%, 80%, 77%, and 60%, respectively.

Conclusion: The overall survival of patients with CTD-PAH was unsatisfactory. Increased CRP level during follow-up indicated that the inflammatory process is crucial in CTD-PAH. Therefore, treatment of PAH and control of inflammation is important in patients with CTD-PAH.