

THE IMPORTANCE OF ECHOCARDIOGRAPHIC SCREENING FOR PULMONARY ARTERIAL HYPERTENSION IN KOREAN PATIENTS WITH SYSTEMIC SCLEROSIS

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Recognition of pulmonary arterial hypertension (PAH) associated with systemic sclerosis (SSc) (SSc-PAH) is very important, because SSc-PAH is highly prevalent and is a leading cause of mortality in patients with SSc.^{1,2)} Despite the use of novel therapeutic strategies, response to PAH-targeted therapy is still insufficient in SSc-PAH, which is associated with poor prognosis in SSc-PAH.³⁾ Indeed, it has been reported that PAH develops in 12% of patients with SSc, and about 50% of SSc patients die within three years of SSc-PAH diagnosis.⁴⁾ Initially, there have been argues that early diagnosis and management of SSc-PAH can improve patients' outcomes,⁵⁾ but recent clinical trials and cohort studies have clearly demonstrated the beneficial effect of early treatment for SSc-PAH.^{6,7)} Therefore, current guideline recommends annual echocardiographic screening for the detection of SSc-PAH.⁸⁾ Right heart catheterization (RHC) is required for patients in whom the results of echocardiography are suggestive of PAH, to confirm diagnosis.⁸⁾

In this issue of the Journal of Cardiovascular Ultrasound, Yoo et al.⁹⁾ performed echocardiography and RHC for the evaluation of PAH in 37 adult Korean patients with SSc. The authors showed that the prevalence of SSc-PAH in study patients was 21.6% in echocardiography and 13.5% in RHC. This SSc-PAH prevalence is similar to prior studies performed in other countries.^{4,10)} The result of this study is meaningful and deserves clinical attention, because this is the first study primarily focused on the prevalence of SSc-PAH in Korea using RHC. Despite of clinical importance of SSc-PAH, there

has been limited data assessing the prevalence of SSc-PAH in Korean patients. To understand and treat a disease properly, it is critical to know the exact prevalence of the disease and to grasp what proportion of patients affected by the disease. Of course, it is obvious that this single center study with small number of subject cannot reflect exact prevalence of SSc-PAH in entire Korea. However, the prevalence can be assumed to be similar as long as significant regional disparities don't exist, because all SSc patients in study hospital were tried to be investigated. Another finding in this study that cannot be ignored is that two patients without any chest symptoms had PAH in echocardiography. This result emphasizes the importance of SSc-PAH screening even in patients without symptoms. Fortunately, recent Korean insurance organization has begun to extend insurance coverage of the use of echocardiography. With this opportunity, more active screening for SSc-PAH should be encouraged, and multi-center studies with a large sample size should be performed to confirm findings of this study. Additionally, studies specifying the characteristics of patients with PAH compared to those without PAH in SSc patients also provide us clinically valuable information, and should be performed. In conclusions, clinicians should recognize the importance of early treatment of SSc-PAH, and make an effort for the early detection of SSc-PAH by the use of useful screening tools such as echocardiography.

REFERENCES

- Hachulla E, Gressin V, Guillevin L, Carpentier P, Diot E, Sibilia J, Kahan A, Cabane J, Francès C, Launay D, Mouthon L, Allanore Y, Tiev KP, Clerson P, de Groote P, Humbert M. *Early detection of pulmonary arterial hypertension in systemic sclerosis: a French nationwide prospective*

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- multicenter study. Arthritis Rheum* 2005;52:3792-800.
2. Steen VD, Medsger TA. *Changes in causes of death in systemic sclerosis, 1972-2002. Ann Rheum Dis* 2007;66:940-4.
 3. Fisher MR, Mathai SC, Champion HC, Girgis RE, Houston-Harris T, Hummers L, Krishnan JA, Wigley F, Hassoun PM. *Clinical differences between idiopathic and scleroderma-related pulmonary hypertension. Arthritis Rheum* 2006;54:3043-50.
 4. Chaisson NF, Hassoun PM. *Systemic sclerosis-associated pulmonary arterial hypertension. Chest* 2013;144:1346-56.
 5. Williams MH, Das C, Handler CE, Akram MR, Davar J, Denton CP, Smith CJ, Black CM, Coghlan JG. *Systemic sclerosis associated pulmonary hypertension: improved survival in the current era. Heart* 2006;92:926-32.
 6. Galiè N, Rubin LJ, Hooper M, Jansa P, Al-Hiti H, Meyer G, Chiossi E, Kusic-Pajic A, Simonneau G. *Treatment of patients with mildly symptomatic pulmonary arterial hypertension with bosentan (EARLY study): a double-blind, randomised controlled trial. Lancet* 2008;371:2093-100.
 7. Humbert M, Yaici A, de Groote P, Montani D, Sitbon O, Launay D, Gressin V, Guillemin L, Clerson P, Simonneau G, Hachulla E. *Screening for pulmonary arterial hypertension in patients with systemic sclerosis: clinical characteristics at diagnosis and long-term survival. Arthritis Rheum* 2011;63:3522-30.
 8. Galiè N, Hooper MM, Humbert M, Torbicki A, Vachiery JL, Barbera JA, Beghetti M, Corris P, Gaine S, Gibbs JS, Gomez-Sanchez MA, Jondeau G, Klepetko W, Opitz C, Peacock A, Rubin L, Zellweger M, Simonneau G; ESC Committee for Practice Guidelines (CPG). *Guidelines for the diagnosis and treatment of pulmonary hypertension: the Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS), endorsed by the International Society of Heart and Lung Transplantation (ISHLT). Eur Heart J* 2009;30:2493-537.
 9. Yoo SJ, Park JH, Park Y, Lee JH, Sun BJ, Kim J, Yoo IS, Shim SC, Kang SW. *Prevalence of pulmonary arterial hypertension in Korean adult patients with systemic sclerosis: result of a pilot echocardiographic screening study. J Cardiovasc Ultrasound* 2016;24:312-6.
 10. Mukerjee D, St George D, Coleiro B, Knight C, Denton CP, Davar J, Black CM, Coghlan JG. *Prevalence and outcome in systemic sclerosis associated pulmonary arterial hypertension: application of a registry approach. Ann Rheum Dis* 2003;62:1088-93.