Pulmonary Arterial Hypertension in Thai Patients with Systemic Sclerosis

Suparaporn Wangkaew MD*, Nuntana Kasitanon MD*,
Arintaya Phrommintikul MD**, Waraporn Sukitawut MD*,
Ramjai Wichainun MD*, Worawit Louthrenoo MD*

** Division of Rheumatology, Department of Internal Medicine, Faculty of Medicine, Chiang Mai University, Chiang Mai*
*** Division of Cardiology, Department of Internal Medicine, Faculty of Medicine, Chiang Mai University, Chiang Mai*

Objective: *Study the clinical features of pulmonary arterial hypertension (PAH) in Thai patients with systemic sclerosis (SSc), and compare these features between those with limited (lc) SSc and diffuse (dc) SSc.*
Material and Method: *The medical records of SSc patients attending the Division of Rheumatology, Chiang Mai University were reviewed. PAH was defined by pulmonary arterial systolic pressure (PASP) > 35 mmHg, determined by Doppler echocardiography.*
Results: *Among 275 patients with SSc, 66 had Doppler echocardiography measurement. Thirty-nine patients (59.1%) had PAH. Among the PAH-SSc patients, 36 (92.3%) presented with dyspnea on exertion, and 37 (94.8%) were in a New York Heart Association functional class of II and III. Twenty-four of 39 patients (61.5%) had interstitial lung disease. Diffuse SSc patients had a significantly higher proportion of males, and shorter disease duration between SSc and PAH diagnosis than lcSSc patients.*
Conclusion: *PAH was not uncommon in Thai patients with SSc. Interstitial lung disease might have been the cause associated with over half of these cases. Annual routine Doppler echocardiography screening for PAH in patients with SSc may detect preclinical PAH, and lead to early management and improved functional outcome.*

Keywords: *Scleroderma, Systemic sclerosis, Pulmonary arterial hypertension*

J Med Assoc Thai 2008; 91 (2): 166-72
Full text. e-Journal: *http://www.medassocthai.org/journal*

A recent consensus categorized pulmonary arterial hypertension (PAH) into multiple groups, including idiopathic PAH (IPAH), familial PAH (FPAH), and PAH associated with other diseases[1]. The presence of PAH is associated with significant mortality and morbidity in patients with connective tissue disease, especially PAH associated with systemic sclerosis (PAH-SSc), which is the connective tissue disease mostly associated with PAH[2-4].

Systemic sclerosis (SSc) is a systemic autoimmune disease, which involves mainly the skin, heart, renal, pulmonary, and gastrointestinal system. SSc is classified into two patterns; limited systemic sclerosis (lcSSc) and diffuse systemic sclerosis (dcSSc), according to the extent of skin involvement. In dcSSc, patients may have earlier organ involvement in the renal, heart, and pulmonary system than in the limited form, which may culminate in time. However, PAH occurs most commonly in lcSSc, or CREST syndrome (Calcinosis, Raynaud's phenomenon, Esophageal dysmotility, Sclerodactyle, Telangiectasia), as in a previous study[5]. The reported prevalence of PAH-SSc has been between 13 and 37.3%, using the echocardiographic measurement of pulmonary arterial pressure[6-14].

SSc is not an uncommon connective tissue disease. Both SSc and PAH pose a significant impact in the patient's quality of life and morbidity. The present report aimed to study the clinical features of PAH-SSc in Thai patients, and compare the clinical findings between the limited and diffuse disease.

Correspondence to : Louthrenoo W, Division of Rheumatology, Department of Internal Medicine, Faculty of Medicine, Chiang Mai University, Chiang Mai 50200, Thailand. Phone: 053-946-449, Fax: 053-357-959, E-mail: wlouthre@mail.med.cmu.ac.th

Material and Method

All adult patients, who were diagnosed SSc based on the American College of Rheumatology criteria[15], and attended the outpatient Rheumatology Clinic at Chiang Mai University Hospital from January 1994 to December 2006, were identified and their clinical records reviewed. Patients with SSc and at least one of the following symptoms, e.g., progressive exertional dyspnea, nonproductive cough, fatigue, chest pain, palpitation, hemoptysis, dizziness or syncope, were referred to cardiologists for echocardiography in order to determine PAH.

Echocardiographic study was performed using a two-dimensional, M mode Doppler echocardiograph. The measurement was performed at rest using a Sonos 5500 (Hewlett Packard, Andover, Massachusetts, USA) equipped with 2.5-3.5 MHz transducers according to the recommendations of the American Society of Echocardiography. Doppler echocardiographic recording in an apical 4-chamber or parasternal short axis view was performed to estimate PASP. Simplified Bernoulli equation was used to estimate peak PASP[16,17]. To estimate right atrial pressure, 10 mmHg was added to the tricuspid pressure gradient to calculate right ventricular systolic pressure or peak pulmonary arterial systolic pressure (PASP) in the absence of right ventricular outflow tract obstruction. PAH was defined as a PASP > 35 mmHg[18]. Only patients with PASP > 35 mmHg were included in the present study. If the patients had several echocardiographic assessments, only the first record was used for analysis. The degree of PAH was classified as mild to moderate, and severe, when PASP pressure was > 35 to 60 mmHg, and > 60 mmHg, respectively[9].

Patients were excluded from the present study if they were less than 16 years old, had a diagnosis of overlap syndrome, or were diagnosed with mixed connective tissue disease. In addition, patients who had signs of clinical heart failure, evidence of left sided heart disease (left ventricular ejection fraction: LVEF < 50%), significant valvular heart disease, congenital heart disease, significant obstructive pulmonary disease, obstructive sleep apnea, portal hypertension, or chronic thromboembolic disease were excluded from the present study. Patients testing positive for antibodies to the human immunodeficiency virus, or had a history of anorexigen use, or other disease known to relate with PAH were also excluded from analysis.

The lcSSc was defined as skin involvement distal to elbows and knees with or without facial involvement. The dcSSc was defined as skin tightening proximal to elbows and knees or truncal involvement. Disease duration was defined as the interval from the date of scleroderma diagnosis to that of PAH diagnosis by Doppler echocardiography.

Clinical records of PAH patients were reviewed to identify the demographic data, clinical manifestations at PAH diagnosis, echocardiogram reports, New York Heart Association (NYHA) functional class, high-resolution computed tomography (HRCT) of lungs reports, laboratory investigations (hemoglobin, serum creatinine, serum albumin), and treatment. HRCT was performed only in patients suspected of having ILD, and those who could afford an HRCT study or obtain financial reimbursement from the government. Interstitial lung disease (ILD) was defined by the presence of definite interstitial infiltration of the lungs in a chest radiograph or the presence of interstitial infiltration with ground glass appearance in HCRT. The present study was approved by the institutional research ethic committee.

Statistical analysis

Continuous variables were described as mean and standard deviation (SD). Categorical variables were described as percentages. Comparisons were made using the Student's t-test for parametric continuous variables, Mann-Whitney U test for nonparametric continuous variables and Chi-square or Fisher's exact test for qualitative variables**.** A p-value of < 0.05 was considered statistically significant. All statistical analyses were performed using the SPSS program statistical software package, version 11.5 for Windows (SPSS Inc, Chicago, Illinois, USA).

Results

Of 275 SSc patients attending the Rheumatology Clinic, 66 (13 males, 53 females, mean age of 47 $\pm$ 14.7years) had Doppler echocardiography measurement. Twenty seven out of 66 patients were excluded from the present study. The reasons for exclusion were estimated-PASP $\leq$ 35 mmHg in 24 patients, significant valvular heart disease in two patients, and significantly impaired LV function in 1 patient. The remaining 39 (59.1%) patients (13 males, 26 females) with PAH had a mean age at SSc diagnosis of 48.5 $\pm$ 14.7 years, mean age at PAH diagnosis of 52.7 $\pm$ 12.2 years, and mean disease duration of 3.2 $\pm$ 2.9 years. There were 20 patients with lcSSc, and 19 with dcSSc. Their mean PASP was 50.9 $\pm$ 15.4 mmHg. Thirty patients (76.9%) had mild to moderate PAH (PASP > 35 to 60 mmHg) and 9 (23.1%) had severe PAH (PASP > 60 mmHg). Among

the PAH-SSc patients, 11 (28.2%) had impaired RV function, 17 (43.6%) had RV enlargement, 10 (25.6%) had RA enlargement, and 10 (25.6%) had systolic D-shape LV (Table 1).

All patients had at least one clinical symptom at PAH diagnosis including progressive exertional dyspnea in 36 (92.3%) cases, nonproductive cough in 21 (55.8%), fatigue in 4 (10.2%), chest pain in 3 (7.7%), and 1 (3.3%) case of each presenting with palpitation, hemoptysis, and syncope. At the time of PAH diagnosis, patients had NYHA class II in 13 (33%) cases, class III in 24 (61.5%), and class IV in 2 (5.1%) (Table 1).

Twenty-four (61.5%) of 39 patients [lcSSc in 13 (54.2%), and dsSSc in 11 (45.8%)] had diffuse interstitial change on chest radiographs. Of these, HRCT was performed in 17 patients, but the results were available only in 15 patients. There were usual interstitial pneumonitis (UIP) in 8 (53.5%) cases, lung fibrosis in 6 (40%), and bronchiolitis obliterans with organizing pneumonia (BOOP) in 1 (6.5%) case. Twenty-three patients were treated with oral cyclophosphamide (50 mg/day) and low dose prednisolone (5.7 $\pm$ 5.8 mg/day) for ILD. One patient who did not receive the treatment was 88 years old, dsSSc, and had multiple medical problems.

PAH was treated with calcium channel blocker in 33 (84.6%) cases, and prostacyclin analogue in 15 (38.5%). Warfarin was administered to 12 (30.8%) patients. Of all PAH patients, the mean duration of follow up was 4.2 $\pm$ 3.4 years, and two patients died, one from progressive RV failure, and the other from sepsis.

A comparison of clinical variables in PAH-SSc patients with limited and diffuse disease is shown in Table 1. There was no statistically significant difference in clinical variables among the two subtypes, except that the proportion of males was significantly higher (57.9% vs. 10.0%; p = 0.005), and disease duration between SSc and PAH diagnosis shorter in dcSSc patients (2.2 $\pm$ 2.2 vs. 4.3 $\pm$ 3.2 years; p = 0.03) than in lcSSc patients.

Discussion

In the present study, the authors found that 39 of 66 (59.1%) SSc patients, with echocardiography

Table 1. The clinical characteristics of lcSSc compared with dcSSc patients with PAH

Characteristics	Limited SSc (n = 20)	Diffuse SSc (n = 19)
Sex (M:F)	2/18	11/8*
Mean age at SSc diagnosis (yrs)	45.6 $\pm$ 9.0	51.5 $\pm$ 18.7
Mean age at PAH diagnosis (yrs)	49.8 $\pm$ 8.4	55.7 $\pm$ 14.9
Mean Disease duration (yrs)	4.3 $\pm$ 3.1	2.2 $\pm$ 2.2**
NYHA class at PAH diagnosis, (No)		
II	6	
III	12	12
IV	2	0
Laboratory investigations		
Mean hemoglobin concentration (g/dl)	11.9 $\pm$ 1.7	11.7 $\pm$ 2.3
Mean serum creatinine (mg/dl)	1.0 $\pm$ 0.7	0.9 $\pm$ 0.5
Mean serum albumin (mg/dl)	3.6 $\pm$ 0.4	3.6 $\pm$ 0.5
Echocardiography findings		
Mean PASP (mmHg)	50.8 $\pm$ 15.3	51.0 $\pm$ 15.9
Impaired RV function, No (%)	8 (40.0)	3 (15.8)
RA enlargement, No (%)	4 (20)	6 (31.6)
RV enlargement, No (%)	9 (45.0)	8 (42.1)
Systolic D-shape LV, No (%)	5 (25.0)	5 (26.3)
Interstitial infiltration on CXR, No (%)	13 (65.0)	11 (57.9)
Interstitial infiltration on HRCT, No (%)	10 (50.0)	5 (26.3)
Treatment with oral cyclophosphamide, No (%)	13(65.0)	10 (52.6)
Mean PaO2 at PAH diagnosis	70.5 $\pm$ 28.9	72.4 $\pm$ 18.6

* p = 0.005, ** p = 0.031

measurement, had PAH. All patients had at least 1 clinical symptom at the time of PAH diagnosis, with dyspnea on exertion (93.2%), and non productive cough (55.8%) being the two most common clinical presentations. The high prevalence of PAH-SSc in this study was higher than the range of 13 to 37.3% reported by previous retrospective studies[6-14], which diagnosed by using 2-dimentional echocardiography with a variety of RVSP cutoffs ranging from 35 to 45 mmHg. These studies were performed in both asymptomatic and symptomatic patients. However, a PAH prevalence of 40% was found in symptomatic patients in the prospective part of one study[9]. Ninety five percent of the patients had an NYHA of class II and III at the time of PAH diagnosis. Thus, patients usually had functional class II or III when PAH was detected. This should lead to more awareness from physicians for the early detection of PAH in SSc patients.

The relatively high prevalence of PAH in the present study was due to a selection bias, as the authors performed an echocardiographic study in only symptomatic patients. All of our PAH-SSc patients had at least one clinical symptom, and over half of them presented with an NYHA of at least class III. Over 25% of PAH-SSc patients had other abnormal echocardiographic findings, such as impaired right ventricular function, right atrial enlargement, right ventricular enlargement, or systolic D-shape LV at the time of PAH diagnosis. The presence of abnormal ECG was associated with a poor prognosis, as abnormal ECG findings usually occurred when PAH was well established[19].

When comparing mean pulmonary arterial pressure measurement assessed by right-heart catheterization (RHC); the gold standard for PAH diagnosis, the PAH-SSc prevalence had a reported range of between 11% and 16%[4,8,20], which is lower than the echocardiographic assessment. However, this measurement is more complicated, and not routinely used for PAH diagnosis in our institution. Mukerjee, et al reported that a tricuspid pressure gradient > 45 mmHg, estimated by echocardiogram, had a good correlation with RHC when identifying PAH patients, giving a sensitivity of 47% and specificity of 97%[21]. Echocardiography is less expensive, noninvasive, and more available for determining PAH. Thus, the authors used Doppler echocardiography to assess PAH in the present symptomatic patients. However, the limitation of the echocardiographic study was that PASP could not be calculated if no transtricuspid gradient was detected. This occurred in 27 of 66 of SSc patients who had clinical symptoms, and the physical signs suggested PAH. Therefore, further RHC in these patients may be helpful to determine a more precise prevalence of PAH in this condition. As the aim of the present study was to determine the clinical features of PAH in SSc patients, the authors did not compare the clinical features among those with and without PAH.

Approximately two-thirds of our PAH-SSc patients had interstitial lung disease in the chest radiograph and approximately 70% of them had HRCT. UIP was the major finding in the HRCT followed by lung fibrosis. All except one patient received oral cyclophosphamide and low dose prednisolone for treatment. Although many of these patients showed some subjective clinical improvement such as in cough and dyspnea, unfortunately, the authors did not systematically measure their lung functions. A recent placebo-controlled study showed that cyclophosphamide was superior to placebo in improving lung function, dyspnea, skin thickness score and health related quality of life[22].

Generally, a lung biopsy would be the most specific method in determining interstitial lung disease. However, this procedure is invasive and can have complications. A restrictive lung disease pattern on a pulmonary function test (PFT) and low diffusing capacity for carbon monoxide (DLco) in patients who have lung fibrosis on HRCT would be more helpful when confirming the diagnosis of lung fibrosis. However, PFT was performed in only 20% of the PAH patients in the present study.

In the present study, most clinical characteristics including mean age of SSc diagnosis or PAH diagnosis, clinical presentation, NYHA functional class, laboratory investigations, echocardiography findings, lung fibrosis on HRCT, and PAH treatment were no different between lcSSc and dcSSc in PAH patients. However, there was a larger proportion of males (57.9% vs. 10.0%; $p = 0.005$), and shorter duration between SSc and PAH diagnosis, in dcSSc patients than in lcSSc patients (2.2 ± 2.2 vs. 4.3 ± 3.2 years; $p = 0.03$). An explanation for the latter finding may be because dcSSc patients, who usually have rapid progression of internal organ involvement, tend to have earlier echocar-diography measurement, due to the selection bias. The present results were in contrast to previous studies in that a majority of PAH-SSc patients were females with lcSSc[6,8,9,11,13].

In previous reports, the incidences of ILD and PAH differed between lcSSc and dcSSc[3,8]. Isolated PAH without ILD is usually the major cause of morbidity and mortality in lcSSc[3,8]. On the other hand,

approximately half of PAH-dcSSc patients are associated with lung fibrosis[3]. Chang B, et al[14] found that PAH-SSc patients with combined interstitial lung disease were more likely to have diffuse disease. Pope JE, et al[9], Hachulla E., et al[23], and Steen V[24] reported a higher prevalence of PAH in dcSSc than the previous studies. Steen V[24] categorized PAH-dcSSc patients into two groups. In the first group, patients developed PAH after ILD-induced hypoxia. In the second group, patients had ILD and later developed severe PAH, with more than the proportional degree of lung fibrosis.

The present study was not consistent with the previous studies, which revealed that ILD was usually predominate in the dcSSc group[3,12-14]. The authors found that lcSSc patients had more lung fibrosis on HRCT than dcSSc patients, although this did not show a statistically significant difference. More patients with lcSSc were sent for HRCT than those in the dsSSc group. This might be due to the former's ability to obtain financial reimbursement from the government, and the physicians were suspicious that ILD was the cause of PAH in these patients. However, the reason for the discrepancy in the present findings from previous studies was not clear, but genetic or racial difference may be one possibility of explaining this result.

There were several limitations in the present study. This was a retrospective study; therefore, relevant information might not have been completely recorded. The small sample size in the present study could affect the statistical analysis. The use of echocardiography to determine PAH has some limitations when calculating PASP in patients, with no transtricuspid regurgitation jet detected or poor image. Moreover, there was a selection bias, as only patients with symptoms or abnormal physical findings, which raised suspicion of PAH, were more likely to have echocardiographic and HRCT assessment. Therefore, the true prevalence of PAH-SSc could not be ascertained in the present study. The diagnosis of lung fibrosis was also problematic, as PFT and DLco tests were not routinely performed at our institution. The authors used clinical symptoms, lung physical signs, chest radiograph, HRCT of the lungs, and arterial blood gas to support the diagnosis of interstitial lung disease and lung fibrosis. Therefore, a further long term prospective study, with a larger sample size and more sensitive diagnostic methods, is required to determine the prevalence in this PAH-SSc population and interstitial lung disease or lung fibrosis.

In conclusion, PAH-SSc in Thai people might not be as uncommon as previously thought. Once the symptoms have developed, patients usually have poor functional outcome and poor cardiac index. Both lcSSc and dcSSc patients have combined lung fibrosis in over one-third of the cases. Annual routine echocardiography screening in both groups of SSc may detect preclinical PAH, leading to early management, and improved clinical outcome in these patients.

References

1. Simonneau G, Galie N, Rubin LJ, Langleben D, Seeger W, Domenighetti G, et al. Clinical classification of pulmonary hypertension. J Am Coll Cardiol 2004; 43: 5S-12S.
2. Ioannidis JP, Vlachoyiannopoulos PG, Haidich AB, Medsger TA Jr, Lucas M, Michet CJ, et al. Mortality in systemic sclerosis: an international meta-analysis of individual patient data. Am J Med 2005; 118: 2-10.
3. Denton CP, Black CM. Pulmonary hypertension in systemic sclerosis. Rheum Dis Clin North Am 2003; 29: 335-49.
4. Humbert M, Sitbon O, Chaouat A, Bertocchi M, Habib G, Gressin V, et al. Pulmonary arterial hypertension in France: results from a national registry. Am J Respir Crit Care Med 2006; 173: 1023-30.
5. Yousem SA. The pulmonary pathologic manifestations of the CREST syndrome. Hum Pathol 1990; 21: 467-74.
6. MacGregor AJ, Canavan R, Knight C, Denton CP, Davar J, Coghlan J, et al. Pulmonary hypertension in systemic sclerosis: risk factors for progression and consequences for survival. Rheumatology (Oxford) 2001; 40: 453-9.
7. Murata I, Kihara H, Shinohara S, Ito K. Echocardiographic evaluation of pulmonary arterial hypertension in patients with progressive systemic sclerosis and related syndromes. Jpn Circ J 1992; 56: 983-91.
8. Launay D, Mouthon L, Hachulla E, Pagnoux C, de Groote P, Remy-Jardin M, et al. Prevalence and characteristics of moderate to severe pulmonary hypertension in systemic sclerosis with and without interstitial lung disease. J Rheumatol 2007; 34: 1005-11.
9. Pope JE, Lee P, Baron M, Dunne J, Smith D, Docherty PS, et al. Prevalence of elevated pulmonary arterial pressures measured by echocardiography in a multicenter study of patients with systemic sclerosis. J Rheumatol 2005; 32: 1273-8.
10. de Azevedo AB, Sampaio-Barros PD, Torres RM, Moreira C. Prevalence of pulmonary hypertension

in systemic sclerosis. Clin Exp Rheumatol 2005; 23: 447-54.
11. Plastiras SC, Karadimitrakis SP, Kampolis C, Moutsopoulos HM, Tzelepis GE. Determinants of pulmonary arterial hypertension in scleroderma. Semin Arthritis Rheum 2007; 36: 392-6.
12. Murata I, Takenaka K, Yoshinoya S, Kikuchi K, Kiuchi T, Tanigawa T, et al. Clinical evaluation of pulmonary hypertension in systemic sclerosis and related disorders. A Doppler echocardiographic study of 135 Japanese patients. Chest 1997; 111: 36-43.
13. Battle RW, Davitt MA, Cooper SM, Buckley LM, Leib ES, Beglin PA, et al. Prevalence of pulmonary hypertension in limited and diffuse scleroderma. Chest 1996; 110: 1515-9.
14. Chang B, Wigley FM, White B, Wise RA. Scleroderma patients with combined pulmonary hypertension and interstitial lung disease. J Rheumatol 2003; 30: 2398-405.
15. Preliminary criteria for the classification of systemic sclerosis (scleroderma). Subcommittee for scleroderma criteria of the American Rheumatism Association Diagnostic and Therapeutic Criteria Committee Arthritis Rheum 1980; 23: 581-90.
16. Yock PG, Popp RL. Noninvasive estimation of right ventricular systolic pressure by Doppler ultrasound in patients with tricuspid regurgitation. Circulation 1984; 70: 657-62.
17. Currie PJ, Seward JB, Chan KL, Fyfe DA, Hagler DJ, Mair DD, et al. Continuous wave Doppler determination of right ventricular pressure: a simultaneous Doppler-catheterization study in 127 patients. J Am Coll Cardiol 1985; 6: 750-6.
18. McGoon MD. Pulmonary hypertension. In: Murphy JR, editor. Mayo clinic cardiology review. 2nd ed. Philadelphia: Lippincott Williams & Wilkins; 2000: 353-71.
19. Geirsson AJ, Wollheim FA, Akesson A. Disease severity of 100 patients with systemic sclerosis over a period of 14 years: using a modified Medsger scale. Ann Rheum Dis 2001; 60: 1117-22.
20. Mukerjee D, St George D, Coleiro B, Knight C, Denton CP, Davar J, et al. Prevalence and outcome in systemic sclerosis associated pulmonary arterial hypertension: application of a registry approach. Ann Rheum Dis 2003; 62: 1088-93.
21. Mukerjee D, St George D, Knight C, Davar J, Wells AU, Du Bois RM, et al. Echocardiography and pulmonary function as screening tests for pulmonary arterial hypertension in systemic sclerosis. Rheumatology (Oxford) 2004; 43: 461-6.
22. Tashkin DP, Elashoff R, Clements PJ, Goldin J, Roth MD, Furst DE, et al. Cyclophosphamide versus placebo in scleroderma lung disease. N Engl J Med 2006; 354: 2655-66.
23. Hachulla E, Gressin V, Guillevin L, Carpentier P, Diot E, Sibilia J, et al. Early detection of pulmonary arterial hypertension in systemic sclerosis: a French nationwide prospective multicenter study. Arthritis Rheum 2005; 52: 3792-800.
24. Steen V. Advancements in diagnosis of pulmonary arterial hypertension in scleroderma. Arthritis Rheum 2005; 52: 3698-700.

ภาวะความดันหลอดเลือดแดงปอดสูงในผู้ป่วยไทยโรคหนังแข็ง

ศุภราภรณ์ วังแก้ว, นันทนา กสิตานนท์, อารินทยา พรหมินธิกุล, วราพร สุขิตาวุธ, แรมใจ วิชัยนันท์, วรวิทย์ เลาห์เรณู

วัตถุประสงค์: *เพื่อศึกษาลักษณะทางคลินิกภาวะความดันหลอดเลือดแดงปอดสูงในผู้ป่วยไทยโรคหนังแข็ง และเปรียบเทียบลักษณะดังกล่าวระหว่างผู้ป่วยโรคหนังแข็งชนิดเฉพาะที่ (limited systemic sclerosis) กับชนิดทั่วไป (diffused systemic sclerosis)*

วัสดุและวิธีการ: *เป็นการศึกษาข้อมูลผู้ป่วยจากเวชระเบียบผู้ป่วยโรคหนังแข็งที่รับการรักษาที่หน่วยโรคข้อ และรูมาติสซั่ม คณะแพทยศาสตร์ มหาวิทยาลัยเชียงใหม่ ภาวะความดันหลอดเลือดแดงปอดสูงหมายถึงภาวะที่ค่าความดันหลอดเลือดแดงปอดช่วงหัวใจบีบตัวมีค่ามากกว่า 35 มม. ปรอทเมื่อทำการตรวจด้วยเครื่องบันทึกภาพหัวใจด้วยคลื่นเสียงความถี่สูง*

ผลการศึกษา: *จากจำนวนผู้ป่วย 275 ราย มีผู้ได้รับการตรวจด้วยเครื่องบันทึกภาพหัวใจด้วยคลื่นเสียงความถี่สูง 66 ราย พบผู้ป่วยมีภาวะความดันหลอดเลือดแดงปอดสูง 39 ราย (ร้อยละ 59.1) จากจำนวน 39 รายนี้ ผู้ป่วย 36 ราย (ร้อยละ 92.3) มีอาการเหนื่อยขณะออกกำลัง และ 37 ราย (ร้อยละ 94.8) มีหน้าที่การทำงานหัวใจอยู่ที่ระดับ 2 และ 3 ตามเกณฑ์ของสมาคมโรคหัวใจแห่งนิวยอร์ก ผู้ป่วย 24 ราย (ร้อยละ 61.5) มีภาวะปอดอักเสบชนิดอินเตอร์สติเชี่ยล (interstitial pneumonitis) ผู้ป่วยโรคหนังแข็งชนิดทั่วไปพบบ่อยกว่าในเพศชาย และมีระยะเวลาจากเริ่มมีอาการหนังแข็งจนถึงการตรวจพบภาวะความดันหลอดเลือดแดงปอดสูงสั้นกว่าเมื่อเปรียบเทียบกับผู้ป่วยโรคหนังแข็งเฉพาะที่*

สรุป: *ภาวะความดันหลอดเลือดแดงปอดสูงพบได้ไม่น้อยในผู้ป่วยไทยโรคหนังแข็ง พบภาวะปอดอักเสบชนิดอินเตอร์สติเชี่ยลซึ่งอาจเป็นสาเหตุได้มากกว่าครึ่งหนึ่งของผู้ป่วยทั้งหมด การตรวจหัวใจด้วยเครื่องบันทึกภาพหัวใจด้วยคลื่นเสียงความถี่สูงเป็นประจำทุกปีในผู้ป่วยกลุ่มนี้จะช่วยในการค้นพบภาวะความดันหลอดเลือดแดงปอดสูงได้ก่อนที่ผู้ป่วยจะมีอาการ ซึ่งจะนำไปสู่การรักษาที่เหมาะสมและให้ผลการรักษาที่ดี*