



## Arthritis News

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### **PULMONARY HYPERTENSION OFTEN GOES UNDIAGNOSED IN PATIENTS WITH CONNECTIVE TISSUE DISEASE**

SAN ANTONIO, TEXAS—Pulmonary hypertension, which is high blood pressure in the lungs, is often not diagnosed in patients with scleroderma and mixed connective tissue disease despite its being a major disease complication and leading cause of death in this patient population, according to research presented this week at the American College of Rheumatology Annual Scientific Meeting in San Antonio, Texas.

Scleroderma and mixed connective tissue disease are two types of rheumatic autoimmune diseases that cause inflammation, damage and scarring in the skin and internal organs, including the lung and its blood vessels. Pulmonary hypertension, a common and devastating complication of these diseases, results in the death of about half of these patients within two to three years after diagnosis. However, the onset of pulmonary hypertension may be without signs and symptoms, like tiredness or shortness of breath. Therefore, diagnosis is often difficult and very little information exists on the prevalence of pulmonary hypertension in patients followed in community practice.

To assess the prevalence of pulmonary hypertension in people with scleroderma and mixed connective tissue disease, researchers studied patients, drawn from 50 medical practices in North America, who had no previous pulmonary hypertension diagnosis. Patients underwent a Doppler echocardiogram (ECHO) of the heart and exercise tolerance questioning. Pulmonary hypertension was considered present if the ECHO determination of estimated right ventricular systolic pressure was greater than 40mmHg. (Systolic pressure, which is the first number on a blood pressure reading, represents the maximum pressure exerted when the heart contracts. A high systolic pressure indicates strain on the blood vessels when the heart is attempting to pump blood into the bloodstream.)

Of the 669 patients who had no previous diagnosis of pulmonary hypertension, 89 (13.3 percent) registered a greater than 40mmHg reading, showing the presence of pulmonary hypertension. This significant number suggests that an echocardiogram evaluation of patients with scleroderma or mixed connective tissue disease should be conducted regularly to detect any need for further evaluation or intervention therapy for pulmonary hypertension. Results also showed that only a minority of these patients ever had a workup to look for pulmonary hypertension.

“Pulmonary hypertension is a silent killer of these patients, difficult to detect by simple bedside examination until the late irreversible stage is present,” said Fredrick M. Wigley, MD, Rheumatology Division, Johns Hopkins University, Baltimore, Maryland, and an investigator in the study. “Patients should be screened regularly because early detection with ECHO technology provides the opportunity to treat with newly available vasoactive medications that potentially can prevent progression to severe life-threatening disease.”

The American College of Rheumatology is the professional organization for rheumatologists and health professionals who share a dedication to healing, preventing disability and curing arthritis and related rheumatic and musculoskeletal diseases. For more information on the ACR’s annual meeting, see [www.rheumatology.org/annual](http://www.rheumatology.org/annual).

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## **The Point Prevalence of Undiagnosed Pulmonary Hypertension (PAH) in Patients with Connective Tissue Disease (CTD) attending Community Based Rheumatology Clinics (UNCOVER Study).**

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**Rationale:** PAH is a major cause of morbidity and mortality among patients with scleroderma (SSc) and mixed connective tissue disease (MCTD), yet physicians often do not detect its presence until the late stages of disease. Most prevalence data come from University or tertiary Centers which are biased toward severe disease; therefore the true prevalence of PAH among patients with CTD is unknown. We sought to determine the point prevalence of undiagnosed PAH in community based rheumatology practices in the USA and Canada.

**Methods:** Fifty practices reviewed the medical records of all their SSc or MCTD patients for a diagnosis of PAH. If the patient had no PAH diagnosis then they entered a prospective study which included a newly performed Doppler echocardiogram (ECHO) to record the estimated right ventricular systolic pressure (ERVSP) and cardiac function; completion of an exercise tolerance questionnaire (scaled in 3 domains from 0–4; 0=severe; 4=no impairment); chart review collection of data on pulmonary function testing (PFT); serology; and history of digital ulcers (DU). PAH was considered present if the ECHO determination of the ERVSP was >40mmHg. Data are reported as means±SD where appropriate. Only chart review was performed for those patients with an existing diagnosis of PAH.

**Results:** 791 of 909 screened patients were evaluable and completed the study; 669 had no previous PAH diagnosis. Of these, 89/669 (13.3%) had an ERVSP >40 mmHg on ECHO. The total prevalence of PAH in the survey was 122 (known) + 89 (newly diagnosed) or 211/791 (26.7%). ECHO data showed 20/89 (22.5%) with ERVSP of ≥ 50 mmHg; 20/89 (22.5%) with increased RV dimension; 25/89 (28.1%) with RA enlargement and an LV ejection fraction of 61.8±10.8%. Patients with ERVSP >40mmHg had decreased exercise tolerance compared to those with <40mmHg (27% compared to 9.5% with at least one grade 1 response, respectively). History of DU was present in 270/791 evaluable patients (34.1%); 79/270 patients with DU (29.3%) had PAH compared to 132/521 without DU but with PAH (25.3%) (p=0.23). Percent predicted DLCO was lower (58.9±24.1) in the patients with PAH compared to those without PAH (71.9±19.7) (p=.005). Percent predicted FVC did not differ significantly between these two groups.

**Conclusion:** A significant number (13.3%) of patients with SSc and MCTD followed in a community rheumatology practice setting have undiagnosed elevated ERVSP consistent with PAH. These patients have ECHO evidence of right ventricular dysfunction, a low DLCO and decreased exercise tolerance. These data suggest that ECHO evaluation of patients with SSc and MCTD is justified to detect patients who may need further evaluation/intervention for PAH

**Disclosure:** F.M. Wigley, Mediquest 5; Genzyme 5; Actelion 2; BIOGEN-Idec 2; Otsuka 2, 5; M.D. Mayes, Actelion Pharmaceuticals 5; J.A. Limia, Actelion Pharmaceuticals 5; D.A. McLain, Amgen 8; Lilly 8; Wyeth 8; Boehringer 8; Ingelheim 8; L. Chapin, None; C. Ward-Able, Actelion Pharmaceuticals 3, 5.

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