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A New Method For The Early Diagnosis Of Pulmonary Hypertension, The Main Cause Of Death In Patients With Scleroderma

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Article Date: 15 Nov 2007 - 0:00 PST

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Researchers at the University of Granada (UGR) from the Systemic Autoimmune Diseases Unit and the Cardiology Service of the Hospital Clinico San Cecilio in Granada have developed an innovative system which will help doctors make the earliest diagnosis of pulmonary hypertension possible, which is the main cause of death for patients suffering from scleroderma, a rare disease which affects approximately 1,200 out of every million people. Until now, the only method to identify pulmonary hypertension was to perform a cardiac catheterization, an invasive technique consisting of the insertion of a hollow and flexible tube (a catheter) through the jugular vein directly into the right side of the heart. This technique is not advisable for all patients because of its complexity and risk.

The new technique was developed by doctor Lourdes Lopez Perez, and directed by lecturers Norberto Ortego Centeno and Jose Luis Callejas Rubio, from the Systemic Autoimmune Diseases Unit of the Medicine Department, at the University of Granada.

They worked in collaboration with the cardiologists Eduardo Moreno Escobar and Pilar Martin de la Fuente. The great advantage of this system, which is based on the ultrasound technique, is that it allows specialists to measure the pulmonary artery systolic pressure so that pulmonary hypertension can be diagnosed earlier.

A destructive disease

Scleroderma is a systemic autoimmune disease, that is to say, it affects various organs of the body, and it is due to an alteration of the immune system. This disease is specially characterised by colour changing of the hands when exposed to cold, which is known as Raynaud's phenomenon, and causes skin swelling from fingers to wrists. Scleroderma is also characterised by damage in the skin of hands, neck and face. This damage resembles little spider veins known as telangiectasias. In addition to this, other organs such as the oesophagus, the kidneys or the lungs may be affected. Lung damage is one of the most significant and serious symptoms, being the leading cause of death among patients suffering from scleroderma. The research team from the University of Granada maintained detailed medical records of each of the 41 patients included in order to accomplish this study. They also included data from both blood and respiratory tests. An echocardiogram was performed on all patients while they were resting and after physical effort on a special bicycle, called a bicycle ergometer.

From the results of this study, it has been concluded that there is a group of patients who present a serious pathological increase of pulmonary artery systolic pressure when exercising. Therefore, these findings are very early evidence of scleroderma. Thanks to this early diagnosis a suitable treatment can be started to modify the scleroderma cycle, thus improving patient's prognosis significantly.

Early diagnosis

Given that pulmonary hypertension is a very serious complication of scleroderma, Lourdes Lopez stresses the importance of diagnosing it as soon as possible in order to foresee the appearance of symptoms ascribed to it, such as breathing difficulty while partaking in physical activity or dyspnea. She also states that using the bicycle ergometer to measure pulmonary artery systolic pressure of the patient after physical activity and performing an echocardiogram are very reliable methods, since they may help to identify scleroderma earlier.

The results of this new research confirms the importance of performing an echocardiogram on patients suffering from scleroderma, since this fatal complication may be indentified before it becomes irreversible. Some of the results of this pioneering research have already been published in prestigious international journals, such as the European Journal of Echocardiography, the official publication of the *European Association of Echocardiography (EAE)*, and Chest, a publication of the American College of Chest Physicians (ACCP).

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