

Original Article

Can We Predict the Severity of Pulmonary Hypertension in Patients With Scleroderma? ☆

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ABSTRACT

Objectives: To describe the clinical–biological characteristics of patients with scleroderma (SS) and pulmonary artery hypertension (PAH).

To establish the relationship between pulmonary functional tests (PFT), Doppler echocardiography (ECHO) and the severity of the PAH.

Material and methods: Retrospective study of patients with SS treated at a tertiary center. All participants received a protocol study, which included a complete analysis and additional tests: ECHO and PFT with carbon monoxide diffusing capacity (DLCO).

Results: Overall, 331 patients were treated, including 68 (20.5%) with PAH. The limited subtype of SS was the most prevalent. The Pearson's correlation coefficient was used for the following variables: FVC-sPAP, FVC/DLCO-sPAP, DLCO-sPAP and TRV-sPAP, showed a significant moderate linear association in the relationship DLCO-sPAP and TRV-sPAP.

29 deaths occurred, with 12 of them related to PAH. The median time between the PAH diagnosis and death was 1.8 years.

Conclusions: The decrease in DLCO and the increase in TRV are negative predictor factor of PAH which, at the same time, means a worsening prognosis for patients with SS.

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¿Podemos predecir la gravedad de la hipertensión arterial pulmonar en pacientes con esclerodermia?

RESUMEN

Objetivos: Describir las características clínico-biológicas de pacientes con esclerodermia (ES) e hipertensión arterial pulmonar (HTAP).

Establecer la relación entre las pruebas funcionales respiratorias (PFR), la ecocardiografía Doppler (eco-Doppler) y la gravedad de la HTAP.

Material y métodos: Estudio retrospectivo de pacientes con diagnóstico de ES seguidos en un centro de tercer nivel. Se les realizó un estudio protocolizado con analítica completa y pruebas complementarias; se estimó la presión arterial pulmonar sistólica (PAPs), la velocidad de reflujo de la válvula tricúspide (VRT), la difusión de monóxido de carbono (DLCO) y la capacidad vital forzada (CVF), por medio de la eco-Doppler y la PFR.

Resultados: Se incluyó a 331 pacientes, de los cuales 68 (20,5%) tenían HTAP. El subtipo de ES más prevalente fue la limitada. Se calculó el coeficiente de correlación de Pearson a las siguientes variables: CVF-PAPs, CVF/DLCO-PAPs, DLCO-PAP y VRT-PAPs, observándose una asociación lineal moderada significativa en la relación DLCO-PAPs y con VRT-PAP.

Se constataron 29 fallecimientos, 12 relacionados a la HTAP. El tiempo medio entre el diagnóstico de HTAP y la muerte fue de 1,8 años.

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Conclusiones: La disminución de la DLCO y el aumento de la VRT son factores predictores de HTAP, que al mismo tiempo condiciona un peor pronóstico en los pacientes con ES.

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Introduction

Pulmonary involvement in patients with SS is considered, after gastrointestinal complications, a frequent visceral complication, with a prevalence of approximately 80%. This complication may be due to diffuse involvement of the pulmonary interstitium or the appearance of pulmonary hypertension (PHT). The latter with a prevalence estimated at 12%–26% depending on diagnostic criteria, from scanning and population estudiada.^{1–3}

PHT is generally considered a late complication of SS. Previous studies found that the TIME between the diagnosis of SS and the beginning of the PHT is between 9.08 ± 6.6^4 and 14 ± 5 years.¹ It is also a major cause of morbidity and mortality in patients, with a median survival of 50% at 12 months of diagnosis.^{3,4}

The definitive diagnosis of PHT is made with right heart catheterization; however, there are another 4 screening measurements used in most studies due to good correlation with the hemodynamic values obtained with cardiac catheterization: first, the measurement of pulmonary artery systolic pressure (sPAP); second, the speed of the tricuspid reflux (VRT), both parameters measured by transthoracic Doppler; third, considered as a predictor, the decrease of the diffusion coefficient of carbon monoxide (DLCO) calculated by means of the RFT, and finally the elevation in serum N-terminal brain natriuretic peptide (N-terminal BNP) prohormone, recently reviewed.^{5–7}

In this paper, we studied a population with SS and PHT with clinical, immunologic, echocardiographic and spirometry parameters, and tried to identify predictors of higher levels of sPAP. We also studied the influence of PHT in the prognosis of these patients.

Materials and Methods

Design

The study included 331 patients with a diagnosis of SS controlled at the Systemic Autoimmune Diseases Unit of the Internal Medicine department, University Hospital Vall d'Hebron, until 2009. Of these patients, we studied 68 patients who met PHT criteria by duplex Doppler considering PHT sPAP values above 35 mmHg.

We divided the population according to clinical characteristics in subtypes of SS (limited, diffuse and no SS). These patients underwent spirometry with DLCO and FVC measurements, a computed tomography (CT), an immunological profile and capillaroscopy following a study protocol used in the department.⁸

Two patterns were considered for capillaroscopy: slow, characterized by the presence of megacapillaries, no hair loss, and a current pattern for important⁹ hair loss.

Statistical Analysis

Statistical analysis was performed using SPSS 11.5 for Windows. The association between variables was performed using the Pearson correlation coefficient and results were considered statistically significant if presenting *P* values of less than .05.

Results

331 patients with SS were included in the study, of which 68 (20.5%) had sPAP > 35 mmHg. The mean age of patients was

Table 1

Clinical, Immunological and Complementary Test Characteristics in Patients With SS and PHT.

	Diffuse	Limited	No Scleroderma
Number of patients	16/68 (24%)	41/68 (60%)	11/68 (16%)
Clinical characteristics			
Raynaud's phenomenon	12/16 (75%)	38/41 (92%)	10/11 (91%)
Capillaroscopy pattern			
Slow	10/16 (62%)	36/41 (87%)	9/11 (81%)
Active	5/16 (32%)	5/41 (13%)	0
Undefined	1/16 (6%)	0	2/11 (18%)
Presence of ulcers	15/16 (93%)	30/41 (73%)	1/11 (9%)
Immunological			
ACA (+)	3/16 (19%)	18/38 (47%)	8/10 (72%)
Scl70 (+)	9/16 (67%)	3/38 (8%)	1/11 (9%)
RFT			
DLCO (media)	55% (n=13)	48% (n=29)	53% (n=7)
CVF (media)	51% (n=14)	63% (n=36)	61% (n=10)
TAC			
Interstitial affection	12/16 (75%)	16/32 (50%)	2/10 (20%)
No interstitial affection	4/16 (25%)	16/32 (50%)	8/10 (80%)
echo-Doppler			
Mean sPAP (mmHg)	62 (n=16)	58 (n=41)	64 (n=11)
Mean VRT (m/s)	2.38 (n=12)	2.33 (n=27)	2.24 (n=9)

RFT: respiratory function tests; DLCO: carbon monoxide diffusion; CVF: forced vital capacity; PAP: pulmonary arterial pressure; ACA: anticentromere antibodies; Scl70: anti sclera 70 antibodies; echo-Doppler: Doppler echocardiography; sPAP: systolic pulmonary arterial pressure; VRT: tricuspid valve reflux speed; n: number of patients with available test results.

56.3 ± 14.4 years), 61 were women (90%) and 7 men (10%), with a 9:1 ratio.

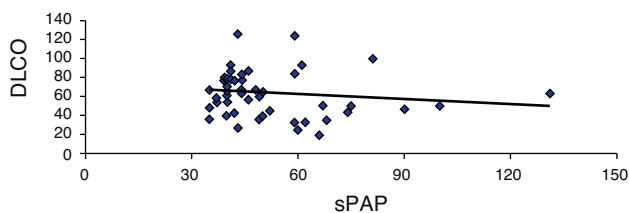
Raynaud's phenomenon was the first most common manifestation of SS in all subtypes studied. The average time from the first manifestation of the disease until the diagnosis of the same was 9.5 ± 11 years. Clinical and immunological characteristics of the study population are shown in Table 1.

The average sPAP by ECHO at rest was 54.9 ± 18 mmHg, while the mean VRT was 3.3 m/s. With respect to respiratory function tests, we found a mean DLCO of $45.7\% \pm 34\%$ predicted, a FVC of $59.5\% \pm 27.8\%$ predicted, and ratio of the FVC/DLCO of 1.3 ± 0.6 .

Using chest CT we found interstitial disease in 52% (30/58) of patients, and 28% (16/58) of patients had a limited form.

We calculated the Pearson correlation coefficient with the following variables FVC-sPAP ($r = -0.040$, $P = .373$), FVC/DLCO-sPAP ($r = 0.150$, $P = .149$), DLCO-sPAP ($r = -0.282$, $P = .01$) and VRT-sPAP ($r = 0.658$, $P = .0001$), demonstrating a linear association of moderate significance in relation to these 2 last variables (Figs. 1 and 2).

We also analyzed other variables such as the presence of distal ulcers ($P = .184$), the active capillaroscopy pattern ($P = .241$), the



$r = -0.282$, $P = .01$

Fig. 1. Correlation between sPAP and DLCO (n=49). $r = -0.282$, $P = .01$.

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