



Doppler Echocardiography Overestimates Pulmonary Hypertension in Patients with Scleroderma Interstitial Lung Disease

Kevin M Chan¹, Elena Tishkowski², Ann J Impens², Tammy Ojo¹, Vallerie V McLaughlin³ and James R Seibold²

Department of Internal Medicine, ¹Division of Pulmonary and Critical Care Medicine, ²Division of Rheumatology, Scleroderma Program, and ³Division of Cardiovascular Medicine, University of Michigan Health Systems, Ann Arbor, MI, USA

Introduction

- Lung disease is the leading cause of mortality and late disease morbidity in systemic sclerosis
- Pulmonary hypertension (PH) affects between 8 and 10% of patients with scleroderma and is associated with a poor prognosis
- Scleroderma patients with combined interstitial lung disease (SILD) and PH are a distinct subpopulation often excluded or under-represented in clinical trials
- Doppler echocardiography (DE) is commonly used to screen for PH in this population

Purpose

- Determine the utility and accuracy of DE to detect PH in patients with SILD

Methods

- Potential participants in a Phase IV clinical trial evaluating an inhaled medication in patients with PH and SILD were reviewed
- SILD was determined by HRCT of the chest, FVC < 70% predicted and DLCO < 60% predicted
- DE measurement of the pulmonary artery systolic pressure (PASP) was determined by tricuspid jet velocity
- Spirometry, diffusing capacity, six minute walk testing and right heart catheterization were required for evaluation
- PH by DE was defined as a PASP of > 40 mmHg
- PH by right heart catheterization (RHC) was defined as a mean pulmonary artery pressure (mPAP) > 25 mmHg at rest
- The accuracy of DE to detect PH when compared to RHC was analyzed
- Demographics and outcome results are expressed as mean ± standard deviation.
- Comparative analyses were performed using a non-parametric t-test, the Mann-Whitney Test

Results

- 13 patients met criteria for review
- 8 patients had PH defined by DE
- 8 patients had PH defined by right heart catheterization

Demographics

	PH by RHC N = 8	No PH by RHC N = 5	p-value
Gender (Female)	7(88%)	4 (80%)	p=0.7
Age (mean±SD)	56 ±7	49 ±11	p=0.2
PSS Type DcSSc/LcSSc	7 / 1	1 / 4	p=0.02
HRCT Diag NSIP/UIP	7 / 1	3 / 2	p=0.3
FVC % Predicted	57 ± 14	49 ± 10	p=0.55
DLCO % Predicted	33 ± 11	22 ± 7	p=0.07
FVC/DLCO	1.82 ± 0.6	2.57 ± 1.45	p=0.46
6MW distance (meters)	332 ± 109	295 ± 75	p=0.53
Difference SpO2 fall after exercise (%)	8.86 ± 5	3.4 ± 4	p=0.06

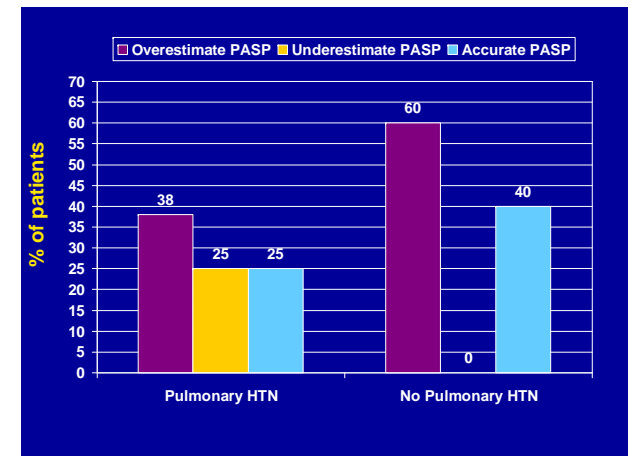
Hemodynamics

	PH by RHC N = 8	No PH by RHC N = 5	p-value
RHC PASP (mmHg)	53 ±19	32 ±6	p=0.008
mPAP(mmHg)	34 ± 11	21 ± 3	p=0.003
PVR (WU)	4.9 ± 3	2.5 ± 0.7	p=0.03
Echo PASP estimate (mmHg)	43 ± 10	44 ± 20	p=0.9
PH by DE	5 (63%)	3 (60%)	p=0.9

Ability of DE to predict PH in SILD

- DE sensitivity, specificity, positive predictive value and negative predictive value for determining PH in patients with SILD were 63%, 40%, 63% and 40%, respectively

Accuracy of DE PASP estimate in SILD



Conclusion

- Doppler echocardiography has a poor discriminative ability to predict the presence of PH in patients with SILD
- DE can overestimate PASP in up to 60% of SILD patients without PH
- The magnitude of change in oxygen desaturation after 6 minute walk testing may assist in predicting PH in patients with SILD

Clinical Implications

- Right heart catheterization should be performed in all patients with SILD when PH is suspected and initiation of lifelong medical therapy is considered

Disclosure Information:
Grant monies: Actelion
Product/Research Disclosure Information: KMC None