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Screening Algorithm for Pulmonary Hypertension in Systemic Sclerosis – Comparison of Predictive Accuracy of Three Algorithms

Vivek Nagaraja¹, Scott H. Visovatti², Heather Gladue³, Veronica J. Berrocal⁴, Jennifer Serrano⁵, Vallerie McLaughlin² and Dinesh Khanna⁶, ¹Department of Medicine [Division of Rheumatology], University of Toledo, Toledo, OH, ²Internal Medicine, Division of Cardiology, University of Michigan, Ann Arbor, MI, ³Rheumatology, Emory University, Atlanta, GA, ⁴Div of Rheumatology, University of Michigan, Ann Arbor, MI, ⁵University of Michigan, Ann Arbor, MI, ⁶Division of Rheumatology, University of Michigan, Ann Arbor, MI

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Session Type: ACR Concurrent Abstract Session

Session Time: 2:30PM-4:00PM

Background/Purpose: Pulmonary arterial hypertension (PAH) is the leading cause of mortality in systemic sclerosis (SSc), and is associated with a 3-year survival of approximately 50%. Early screening for SSc-PAH may improve survival. We compared the predictive accuracy of three recently published screening algorithms – DETECT 2013, Australian Scleroderma Interest Group (ASIG) 2012, Cochin risk prediction score (RPS) 2011 – for SSc-PAH.

Methods: We included consecutive SSc patients with suspected PAH undergoing right heart catheterization (RHC). The inclusion criteria were based on 2013 recommendations for screening PAH (Khanna D. Arthritis Rheum. 2013). The three screening models were applied to each patient. For each model, contingency table analysis was used to determine sensitivity, specificity, and positive (PPV) and negative (NPV) predictive values for PAH [defined as mean pulmonary artery pressure (mPAP) \geq 25, pulmonary capillary wedge pressure (PCWP) \leq 15, and no/mild interstitial lung disease (ILD) on high-resolution CT scan of chest (HRCT), or FVC \geq 70%], WHO group 2 pulmonary hypertension (PH defined as mPAP \geq 25, PCWP $>$ 15, and no / Mild ILD on HRCT, or FVC \geq 70%), and WHO group 3 PH (defined as mPAP \geq 25, PCWP \leq 15, and moderate / severe ILD on HRCT, or FVC $<$ 70%).

Results: Of the 108 patients screened for PAH, 77 met the

recommendations, and 60 patients had the RHC. The prevalence of PAH was 18%. Figure 1 provides a flowchart of patients screened for PAH. There were no significant differences in the baseline clinical characteristics between the PH and non-PH patients. Majority of the patients were females (60% vs 57%), had telangiectasia (70% vs 78%) and about a third of the patients had anticentromere antibody (35% vs 38%). DETECT and ASIG algorithms performed similarly in detecting PAH with sensitivities and NPV of 100 % (Table 1). Approximately 1/3 of patients who met the criteria had PAH (PPV 32-38%). In detecting group-2 PH, DETECT and RPS algorithms had sensitivities and NPV of 100% (Table 1).

Conclusion: In this cohort, the DETECT and ASIG algorithms were comparable in detecting PAH in the SSc patients.

Figure 1: Flowchart of patients screened for PAH

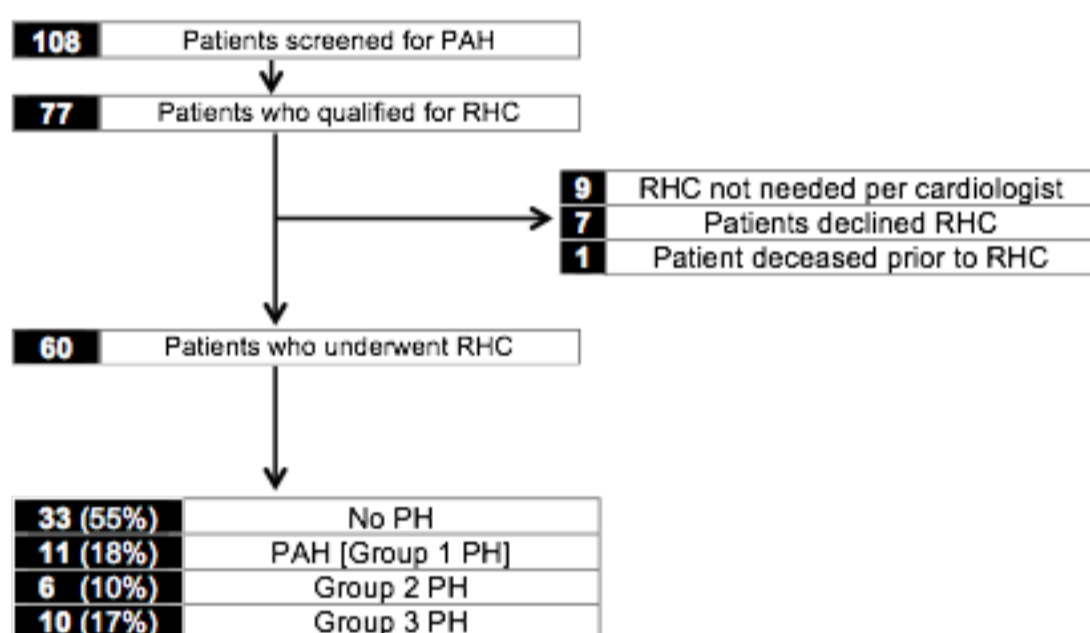


Table 1: Summary of the predictive accuracies (in percentage) of the screening models for PAH in SSc patients

	ASIG			DETECT			RPS		
	PAH	Group-2 PH	Group-3 PH	PAH	Group-2 PH	Group-3 PH	PAH	Group-2 PH	Group-3 PH
Positive	58	47	50	66	60	58	74	73	71
Negative	42	53	50	34	40	42	26	27	29
Sensitivity	100	50	62	100	100	70	91	100	75
Specificity	53	53	53	45	45	45	32	36	30
PPV	37	12	25	38	22	28	32	22	50

NPV	100	89	85	100	100	83	91	100	83
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PPV positive predictive value; NPV negative predictive value; PAH pulmonary arterial hypertension; PH pulmonary hypertension

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