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

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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) > or = 25, pulmonary capillary wedge pressure (PCWP) < or = 15, and no/mild interstitial lung disease (ILD) on high-resolution CT scan of chest (HRCT), or FVC > or = 70%], WHO group 2 pulmonary hypertension (PH defined as mPAP > or = 25, PCWP >15, and no / Mild ILD on HRCT, or FVC > or = 70%), and WHO group 3 PH (defined as mPAP > or = 25, PCWP < or = 15, and moderate / severe ILD on HRCT, or FVC <70%).

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

![Flowchart of patients screened for PAH](image)

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

<table>
<thead>
<tr>
<th></th>
<th>ASIG</th>
<th>DETECT</th>
<th>RPS</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Positive</strong></td>
<td>58</td>
<td>47</td>
<td>50</td>
</tr>
<tr>
<td><strong>Negative</strong></td>
<td>42</td>
<td>53</td>
<td>50</td>
</tr>
<tr>
<td><strong>Sensitivity</strong></td>
<td>100</td>
<td>50</td>
<td>62</td>
</tr>
<tr>
<td><strong>Specificity</strong></td>
<td>53</td>
<td>53</td>
<td>53</td>
</tr>
<tr>
<td><strong>PPV</strong></td>
<td>37</td>
<td>12</td>
<td>25</td>
</tr>
</tbody>
</table>
NPV

PPV positive predictive value; NPV negative predictive value; PAH pulmonary arterial hypertension; PH pulmonary hypertension

Disclosure: V. Nagaraja, None; S. H. Visovatti, None; H. Gladue, None; V. J. Berrocal, None; J. Serrano, None; V. McLaughlin, None; D. Khanna, None.

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