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Background: Antinuclear antibodies (ANA) are serological markers for the presence of rheumatic diseases [1]. Some patients may be patterned in ANA immunofluorescence that resembles an anti-dense-fine-speckled-70 antibody [2]. This is detected less frequently in patients with connective tissue diseases (CTD) and is therefore often used as an exclusion criteria for a CTD [3]. To date, however, it is not clear how the presence of an anti-DSF70 autoantibody will reliably rule out CTDs.

Objectives: To examine the degree to which CTDs can be excluded from either a positive test and whether there are discrepancies between patients with CTDs and a negative anti-DSF70 test and those with CTDs and a positive anti-DSF70 test.

Methods: We analyzed data of 460 patients who were tested for the presence of the DSF70 antibody at the University Hospital Bonn, Germany. Patients were examined with regard to clinical symptoms and signs, type of disease, type of CTD, fulfillment of the classification criteria for the particular CTD, DSF70 antibodies, other autoantibodies and ANA titer. Gold standard for presence of a CTD was the diagnosis of the treating rheumatologist.

In addition to variations between patients with CTD and positive or negative DSF70 antibodies, specificity, sensitivity and positive predictive value were calculated for different ANA titers.

Results: Of the 460 patients, 79.8% were female and 182 were diagnosed with a CTD. 354 patients (77%) were tested negative, 81 (17.6%) positive and 25 (5.5%) had a borderline anti-DSF70 autoantibody result. We observed no CTD in 68.2% of the tested patients with CTD positive anti-DSF70 autoantibody positive tested patients. We identified 21 patients with a positive test result in the absence of a CTD, which accounts for 25.9% of all anti-DSF70 autoantibody positive tested patients. We found no significant differences in DSF70-positive patients with CTD in regard to age, gender, symptoms, clinical signs and also disease-specific antibodies compared to patients with negative DSF70 status and present CTD. Of the 21 DSF70-positive patients with CTD, 12 (57.1%) were tested for the presence of disease-specific autoantibodies. Specific autoantibodies could be detected in all of the tested patients with systemic lupus erythematosus, systemic sclerosis, Sjögren's syndrome and mixed connective tissue disease. DSF70 autoantibodies had a specificity of 86.8%, a sensitivity of 26.9% and a positive predictive value of 68.2% at an ANA titer of ≥1:160, with respect to the absence of a connective tissue disease.

Conclusion: Autoantibodies to DSF70 often occur in CTD patients and are therefore not a valid exclusion criteria for a CTD. We observed no CTD in 68.2% of patients who were tested positive. Thus, in difficult situations, the DSF70 test may be helpful and the additional exclusion of other autoantibodies may render CTD even less probable.

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Table 1. Sensitivity, specificity and positive predictive value of anti-DSF70 autoantibody in different ANA titers (borderline patients excluded)

<table>
<thead>
<tr>
<th>ANA-Titer</th>
<th>No present CTD</th>
<th>Present CTD</th>
<th>Positive predictive value</th>
</tr>
</thead>
<tbody>
<tr>
<td>N=382</td>
<td>152</td>
<td>21</td>
<td>71.2%</td>
</tr>
<tr>
<td>DSF70 positive</td>
<td>21</td>
<td>21</td>
<td></td>
</tr>
<tr>
<td>DSF70 negative</td>
<td>145</td>
<td>144</td>
<td></td>
</tr>
<tr>
<td>Positive predictive value</td>
<td>71.2%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>N=167</td>
<td>45</td>
<td>21</td>
<td>68.2%</td>
</tr>
<tr>
<td>DSF70 positive</td>
<td>21</td>
<td>21</td>
<td></td>
</tr>
<tr>
<td>DSF70 negative</td>
<td>122</td>
<td>138</td>
<td></td>
</tr>
<tr>
<td>Positive predictive value</td>
<td>68.2%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>N=160</td>
<td>47</td>
<td>17</td>
<td>64.6%</td>
</tr>
<tr>
<td>DSF70 positive</td>
<td>17</td>
<td>14</td>
<td></td>
</tr>
<tr>
<td>DSF70 negative</td>
<td>48</td>
<td>88</td>
<td></td>
</tr>
<tr>
<td>Positive predictive value</td>
<td>54.8%</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Table 1. Main clinical features according to different diagnostic criteria.

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>ISG criteria (N=96)</th>
<th>ICRD criteria (N=96)</th>
<th>Expert rheumatologists (N=120)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age, years / (BD)</td>
<td>50 (14.3)</td>
<td>52 (16.8)</td>
<td>51 (15.6)</td>
</tr>
<tr>
<td>Gender, men/women, N (%)</td>
<td>56 (51)</td>
<td>58 (55.8)</td>
<td>56 (55)</td>
</tr>
<tr>
<td>Oral aphthosis, N (%)</td>
<td>113 (94.2)</td>
<td>59 (100)</td>
<td>104 (97.9)</td>
</tr>
<tr>
<td>Genital aphthosis, N (%)</td>
<td>71 (78.5)</td>
<td>46 (78)</td>
<td>71 (74)</td>
</tr>
<tr>
<td>Skin manifestations N (%)</td>
<td>76 (83.3)</td>
<td>52 (86.1)</td>
<td>64 (71.6)</td>
</tr>
<tr>
<td>Gastrointestinal features, N (%)</td>
<td>54 (44.5)</td>
<td>31 (52.5)</td>
<td>50 (52.1)</td>
</tr>
<tr>
<td>Joint manifestations, N (%)</td>
<td>78 (66)</td>
<td>38 (64.4)</td>
<td>62 (64.6)</td>
</tr>
<tr>
<td>Vascular manifestations, N (%)</td>
<td>23 (19.2)</td>
<td>9 (15.2)</td>
<td>20 (21.1)</td>
</tr>
<tr>
<td>Neurological manifestations, N (%)</td>
<td>38 (32.4)</td>
<td>5 (8.7)</td>
<td>37 (34.8)</td>
</tr>
<tr>
<td>Ocular lesions, N (%)</td>
<td>38 (32.4)</td>
<td>5 (8.7)</td>
<td>37 (34.8)</td>
</tr>
<tr>
<td>Neurological manifestations, N (%)</td>
<td>23 (19.2)</td>
<td>9 (15.2)</td>
<td>20 (21.1)</td>
</tr>
<tr>
<td>Gastrointestinal features, N (%)</td>
<td>8 (6.6)</td>
<td>3 (5.1)</td>
<td>5 (5.3)</td>
</tr>
</tbody>
</table>

Objectives: To assess a) the concordance and differences between ISG and ICRD criteria b) sensitivity in diagnosing severe manifestations (ocular, vascular and neurological).

Methods: The study included 120 patients diagnosed with definite or possible BD by expert rheumatologists. They were diagnosed at a well-defined population in Northern Spain between January 1980 and December 2019. The ISG and ICRD diagnostic criteria for BD were applied to all patients and compared among them.

Results: 120 patients (62 men/58 women) were studied. Mean age at diagnosis was 37.6±13.8 years. 59 (49.2%) patients fulfilled ISG criteria and 96 (80%) ICRD criteria. Concordance between both criteria was moderate (Kappa 0.41). ICRD criteria diagnosed more patients with neurological (χ²=49.1, p<0.01), vascular (χ²=56.7, p<0.01) and ocular manifestations (χ²=84.4 p<0.01) (Figure 1).

Disclosure of Interests: Maria-Masi Mockenhaupt: None declared, Ramona Dolscheid-Pommerich: None declared, Charlotte Behning: None declared, Birgit Stoffel-Wagner: None declared, Peter Brossart: None declared, Valentin Schäfer: Speakers bureau: Abbvie, Novartis, BMS, Chugai, Celgene, Medac, Sanofi, Lilly, Hexal, Pfizer, Janssen, Roche, Schierke, Onkowissen, Royal College London, Consultant of: Novartis, Chugai, AbbVie, Celgene, Sanofi, Lilly, Hexali, Pfizer, Amgen, BMS, Roche, Gilead, Medac, Grant/research support from: Novartis, Hexali, Lilly, Roche, Celgene, Universitäts Bonn
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Figure 1. Number of patients with vascular, neurological or ocular manifestations diagnosed with BD by different criteria. Abbreviations: ITRC-ICBD: International Team for the Revision of the International Criteria for BD; ISG: International Study Group for Behçet Disease.