Paediatric rheumatology

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VALIDATION OF THE PEDIATRIC BEHÇET DISEASE CRITERIA (PEDBD): A REAL LIFE CONSENSUS-BASED APPROACH

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Background: Behcet’s syndrome (BS) is an autoimmune disease characterized by a variable vessel vasculitis. In children, BS may start early in life, mimicking other autoinflammatory diseases and making the diagnosis challenging. In the past, several criteria have been created for adult BS classification. In 2015, the first set of BS paediatric classification criteria, the PEDBD, was proposed by an international Expert consensus [1].

Objectives: to perform an external validation of the PEDBD criteria in a cohort of internationally validated paediatric BS patients.

Methods: to perform an external validation of the PEDBD criteria in a cohort of internationally validated paediatric BS patients.

Results: to perform an external validation of the PEDBD criteria in a cohort of internationally validated paediatric BS patients.

Conclusions: to perform an external validation of the PEDBD criteria in a cohort of internationally validated paediatric BS patients.

Table 1

<table>
<thead>
<tr>
<th>Criteria</th>
<th>Sensitivity</th>
<th>Specificity</th>
<th>Accuracy</th>
</tr>
</thead>
<tbody>
<tr>
<td>ISG</td>
<td>0.50</td>
<td>1.00</td>
<td></td>
</tr>
<tr>
<td>ICBD</td>
<td>0.79</td>
<td>0.98</td>
<td>0.95</td>
</tr>
<tr>
<td>PEDBD</td>
<td>0.58</td>
<td>0.99</td>
<td>0.91</td>
</tr>
</tbody>
</table>

Conclusion: the PEDBD criteria were extremely specific but had a lesser sensitivity than ICBD which had a better accuracy. One limitation is that specific monoclonal BS mimics were not included as disease controls, thus the true accuracy of all these criteria may be lower in practice. The complexity of childhood BS suggests that genotyping (incorporating autoinflammatory diseases, BS mimics, and HLA-type) combined with clinical features are likely to yield the most accurate classification criteria, which would require further validation in a larger cohort.


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