Validation of the 2021 EAN/PNS diagnostic criteria for chronic inflammatory demyelinating polyneuropathy

Satoshi Kuwabara , Tomoki Suichi

The first validation studies of the 2021 EAN/PNS criteria for the diagnosis of CIDP have shown the acceptable sensitivity/specificity

Chronic inflammatory demyelinating polyneuropathy (CIDP) is the most common immune-mediated neuropathy. Because of the lack of disease-specific diagnostic biomarkers, the diagnosis depends on combination of clinical, electrodiagnostic, and laboratory/neuroimaging findings, as well as exclusion criteria; and in clinical practice, misdiagnosis is not uncommon.

In 2021, the revised guideline on CIDP was published from the European Academy of Neurology (EAN; formerly European Federation of Neurological Societies, EFNS) and Peripheral Nerve Society (PNS). The 2021 EAN/PNS guideline is an updated version of the previous 2010 EFNS/PNS guideline, that aims to reflect recent advances of electrodiagnosis and peripheral nerve imaging (ultrasound and MRI), and include to treatment response as a supportive criterion. Additionally, the guideline simplified the diagnostic category: among possible, probable and definite CIDP in the 2010 criteria; probable/definite CIDP is combined and termed just ‘CIDP’ in the 2021 criteria.

Two JNPP studies reported the sensitivity and specificity of the 2021 EAN/PNS criteria, and the results are compared with those of the 2010 EFNS/PNS criteria. Doneddu et al described results of an Italian multicentre study based on Italian CIDP registry database (n=330), whereas Rajabally et al reported analyses on a CIDP cohort of a single centre in the UK (N=120). Table 1 summarises the results of the two studies. Both studies provide useful reference data of diagnostic accuracy for CIDP by the 2021 EAN/PNS guideline. The specificity of the criteria appears sufficient, and future studies are required to further increase the diagnostic sensitivity that will lead to timely and appropriate treatment and improvement in outcome of patients with CIDP.

Table 1 The sensitivity/specificity in the two studies

<table>
<thead>
<tr>
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<th>2021 EAN/PNS criteria</th>
<th>2010 EFNS/PNS criteria</th>
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<tbody>
<tr>
<td></td>
<td>Sensitivity</td>
<td>Specificity</td>
</tr>
<tr>
<td>Doneddu et al</td>
<td>74%</td>
<td>77%*</td>
</tr>
<tr>
<td>(n=330) Italian registry</td>
<td></td>
<td></td>
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<tr>
<td>Rajabally et al</td>
<td>83%</td>
<td>94%</td>
</tr>
<tr>
<td>(n=120) single centre</td>
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</tbody>
</table>

*With supportive criteria.

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Contributors SK wrote a draft of the manuscript, and TS revised it.

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Competing interests None declared.

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Editorial commentary

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