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.[1] 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)[2]; 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 *JNNP* 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.[3 4] Doneddu *et al* described results of an Italian multicentre study based on Italian CIDP registry database (n=330),[3] whereas Rajabally *et al* reported analyses on a CIDP cohort of a single centre in the UK (N=120).[4] Table 1 summarises the results of the two studies. Both studies show very high specificity, 98% and 94%, respectively. However, the sensitivity was somewhat different: 74% in the Italian and 83% in the UK studies. The difference is likely to result from the different nature of each primary cohort. The Italian study used the multicentre registry database and selected patients with possible CIDP by the 2010 criteria; whereas in the UK study, only patients with objective treatment response were included. Additionally, electrodiagnostic evaluation was performed using exactly the same protocol in the UK study; objectively defined clinical response to immune therapy strongly suggests the CIDP diagnosis, and this may explain the higher sensitivity. When such supportive criteria were employed, the sensitivity increased from 74% to 77% in the Italian study.

The sensitivity of 77%–83% and the specificity of 94%–98% are acceptable for research and clinical practice in CIDP. However, further higher sensitivity would be ideal, and we suggest that particularly for patients who have suspectable CIDP and do not fulfil the electrodiagnostic criteria, the use of peripheral nerve imaging with ultrasound and/or MRI, which was not widely performed in the two studies, is recommended. Prominent peripheral nerve enlargement is nearly specific to CIDP and Charcot-Marie-Tooth disease type 1 (a demyelinating form). Among supportive criteria for the CIDP diagnosis (CSF protein, nerve hypertrophy, nerve biopsy and treatment response),[2] treatment response significantly contributes to the diagnosis, but this is not obtained before treatment. We think that neuroimaging would increase the sensitivity.

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.[5]

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Table 1 The sensitivity/specificity in the two studies

	2021 EAN/PNS criteria		2010 EFNS/PNS criteria	
	Sensitivity	Specificity	Sensitivity	Specificity
Doneddu *et al*[3] (n=330) Italian registry	74% 77%*	98%	85%	84%
Rajabally *et al*[4] (n=120) single centre	83%	94%	86%	94%

*With supportive criteria.
EAN, European Academy of Neurology; EFNS, European Federation of Neurological Societies; PNS, Peripheral Nerve Society.

Neurology, Graduate School of Medicine, Chiba University, Chiba, Japan

Correspondence to Dr Satoshi Kuwabara, Neurology, Chiba University Graduate School of Medicine, Chiba 260-8670, Japan; kuwabara-s@faculty.chiba-u.jp

ORCID iDs
Satoshi Kuwabara http://orcid.org/0000-0002-4716-8578
Tomoki Suichi http://orcid.org/0000-0002-9575-9444

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