

EUROPEAN GUIDELINE:

Diagnosis and treatment of chronic inflammatory demyelinating polyradiculoneuropathy (CIDP)

A first revision of the consensus guideline on the definition, investigation, and treatment of CIDP was published in 2010. The aim was to develop a second revision of this guideline, using GRADE (Grading of Recommendations, Assessment, Development, and Evaluation) methodology.



DIAGNOSTIC CRITERIA:

- Typical CIDP and CIDP variants (multifocal, focal, distal, motor, or sensory CIDP) are well characterized entities with specific clinical and electrodiagnostic phenotypes
- Based on the strength of the electrodiagnostic evidence for demyelination, two levels of diagnostic certainty are distinguished; CIDP and possible CIDP
- If electrodiagnosis confirms only possible CIDP, fulfilment of 2 supportive criteria (CSF, imaging, nerve biopsy or objective response to treatment) upgrades the diagnosis to CIDP
- Chronic immune sensory polyradiculopathy (CISP) and chronic autoimmune nodopathies are not classified as CIDP variants



THE PRINCIPAL TREATMENT RECOMMENDATIONS ARE:

- Intravenous immunoglobulin (IVIg) or corticosteroids are strongly recommended as initial treatment in typical CIDP and CIDP variants
- Plasma exchange is strongly recommended if IVIg and corticosteroids are ineffective
- IVIg should be considered as the first-line treatment in motor CIDP (Good Practice Point)
- For maintenance treatment, IVIg or subcutaneous immunoglobulin (SCIg) as well as corticosteroids are recommended
- If the maintenance dose of any of these treatments is high, combination treatments or adding an immunosuppressant drug should be considered (Good Practice Point)



Read the full guideline in the European Journal of Neurology: <https://doi.org/10.1111/ene.14959>

This guideline was developed using the GRADE framework.

When using GRADE, panels make strong recommendation when most clinicians and patients would choose the recommended course of action. Weak recommendation indicates that clinicians and patients should consider the recommended course of action, but the final decision should be based on discussion, case-by-case risks and preferences.

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