

How I treat chronic inflammatory demyelinating polyneuropathy podcast

Author: Urvi Desai, MD¹

Author affiliation: ¹ Neurosciences Institute, Atrium Health Wake Forest Baptist, Charlotte, a facility of Carolinas Medical Center, Charlotte, NC, USA

Corresponding author: Dr Urvi Desai (udesai@wakehealth.edu, Urvi.desai@atriumhealth.org)

Supplementary Materials

Table 1

Summary of guideline updates. CIDP, chronic inflammatory demyelinating polyneuropathy; DADS, distal acquired demyelinating symmetric polyneuropathy; LSS, Lewis-Sumner syndrome MADSAM, multifocal acquired demyelinating sensory and motor neuropathy.

	2010 guideline (1)	2021 guideline (2)
Definitions	Typical CIDP Atypical CIDP	Typical CIDP CIDP variants
Clinical diagnostic criteria	Typical CIDP Chronically progressive, stepwise, or recurrent symmetric proximal and distal weakness and sensory dysfunction of all extremities, developing over at least 2 months; cranial nerves may be affected; and absent or reduced tendon reflexes in all extremities Atypical CIDP Predominantly distal (DADS) or Asymmetric [MADSAM/ Lewis–Sumner syndrome] or Focal (e.g., involvement of the brachial or lumbosacral plexus or of one or more peripheral nerves in one upper or lower limb) or Pure motor or Pure sensory (including chronic immune sensory polyradiculopathy affecting the central process of the primary sensory neuron)	Typical CIDP Progressive or relapsing, symmetric, proximal and distal muscle weakness of upper and lower limbs, and sensory involvement of at least two limbs, developing over at least 8 weeks, and absent or reduced tendon reflexes in all limbs CIDP variants Distal CIDP: distal sensory loss and muscle weakness predominantly in lower limbs Multifocal CIDP: sensory loss and muscle weakness in a multifocal pattern, usually asymmetric, upper limb predominant, in more than one limb; although usually affects upper limbs first, lower limbs may become involve later or be affected from the outset Focal CIDP: sensory loss and muscle weakness in only one limb Motor CIDP: motor symptoms and signs without sensory involvement Sensory CIDP: sensory symptoms and signs without motor involvement
Electrophysiological diagnostic criteria	Definite CIDP: Conduction abnormality in two motor nerves Probable CIDP: Less severe conduction abnormality in two motor nerves Possible CIDP: Conduction abnormality in one motor nerve	CIDP: Conduction abnormality in two motor nerves and two sensory nerves Possible CIDP: Conduction abnormality in one motor nerve and one sensory nerve
Supportive diagnostic criteria (imaging, CSF analysis, nerve biopsy, and treatment response)	Probable CIDP + one supportive criteria = Definite CIDP Possible CIDP + two supportive criteria = Definite CIDP	Possible CIDP + two supportive criteria = CIDP

	Possible CIDP + one supportive criteria = Probably CIDP	
--	---	--

References

1. Van den Bergh PY, Hadden RD, Bouche P, Cornblath DR, Hahn A, Illa I, et al. European Federation of Neurological Societies/Peripheral Nerve Society guideline on management of chronic inflammatory demyelinating polyradiculoneuropathy: report of a joint task force of the European Federation of Neurological Societies and the Peripheral Nerve Society - first revision. Eur J Neurol. 2010;17(3):356-63
2. Van den Bergh PYK, van Doorn PA, Hadden RDM, Avau B, Vankrunkelsven P, Allen JA, et al. European Academy of Neurology/Peripheral Nerve Society guideline on diagnosis and treatment of chronic inflammatory demyelinating polyradiculoneuropathy: Report of a joint Task Force-Second revision. J Peripher Nerv Syst. 2021;26(3):242-68.