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Defining Clinical Terms for Managing CIDP

The GBS|CIDP Foundation International's Inaugural Leadership Collaborative

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↘ ABOUT

The GBS|CIDP Foundation International is a global nonprofit supporting individuals and families affected by GBS, CIDP, MMN, and related conditions.

We improve quality of life through support, education, research, and advocacy–providing trusted information, advancing awareness, funding research, and championing policies that benefit our patient community.

The GBS|CIDP Foundation International recently launched a program aimed at solving an urgent problem related to treatment and care of GBS, CIDP, or MMN. The inaugural collaborative brought together the patient community and global key opinion leaders to define key terms for the management of CIDP. Before standardizing these definitions, many stakeholders created their own conflicting definitions, causing confusion among physicians, regulators, and patients.

The Leadership Collaborative process for defining the terms started with a discussion during an international meeting. Then, the Foundation hosted a patient workshop over two days where patients discussed the terms and provided feedback and information on what key opinion leaders should consider when defining the terms. Finally, a group of global CIDP experts met over a weekend to define the terms using a modified Delphi consensus building approach.

↘ PURPOSE

Historically, Key terms used to describe CIDP management have not been uniformly defined. What it means to "respond to treatment," "relapse," or reach "remission" has often been left to individual physicians, leading to patient confusion and differing clinical trial approaches. Therefore, the GBS|CIDP Foundation International sought to standardize these terms.

In addition to helping patients better understand their CIDP status, standardizing these terms is essential for improving clinical trials testing new therapeutics for CIDP.

This toolkit provides a reference guide for patients, healthcare providers, regulators, and researchers, with the goal of improving consistency in how these terms are used.

Experts drew upon available evidence, historical context, and clinical expertise to draft definitions and come to a consensus on definitions for the following terms:

- **Immune-Neuropathy, Evidence of Disease Activity (IN-EDA)**
- **Immune Neuropathy, No Evidence of Disease Activity (IN-NEDA)**
- **Minimal, Partial, Optimal Response to Treatment**
- **Relapse**
- **Possible Relapse**
- **Refractory**
- **Remission**
- **Residual Symptoms**

↘ FULL DEFINITIONS, FOR MEDICAL PROFESSIONAL USE

The Definitions listed below represent the vernacular that was voted upon by the Leadership Collaborative Key Opinion Leader Task Force. This group of CIDP global experts utilized existing evidence and clinical experience to agree upon definitions for these terms relating to the management of CIDP.

These definitions should not be modified when used in conversation or when describing patients' status.

Journal publication of manuscript referencing the process and terms is coming soon.

Term	Definition
IN-EDA	Patient perception of change* AND a change in disability**, impairment***, or both in any period of time. There must be no other condition that better explains change in disability and impairment.
IN-NEDA 1	No patient perception of change* AND either no decline in Disability** OR no decline in Impairment*** for a duration of 6 months or more.
IN-NEDA 2	No patient perception of change* AND no decline in Disability** AND no decline in Impairment*** components for a duration of 6 months or more.

Recommendation for clinical practice and clinical trials:
Qualify IN-EDA or IN-NEDA status as "on treatment" or "off treatment"
Recommendation for clinical trials: target NEDA-2 status

Minimal Response	Patient perception of improvement AND a minimum change on at least one outcome measure [Standard/low MCID threshold: INCAT ≥ 1 point, I-RODS ≥ 4 centile points, grip strength ≥ 8 kPa (vigorimeter), grip strength ≥ 10% 3-day average, 20% or 4 kg (Jamar dynamometer), MRC-SS ≥ 2 points], assessed within 12 weeks of treatment initiation or change.

* Patient Global Impression of Change (PGIC): ≥1 point change

** Disability: : I-RODS ≥4-point (centile) change or INCAT ≥ 1-point change

***Impairment: Grip strength vigorimeter ≥8 kPa, Jamar dynamometer ≥ 4 kg, Jamar dynamometer ≥ 10% (requires 3 consecutive day average), Jamar dynamometer ≥ 20%, or MRC 60 sum score ≥ 2-point change

Partial Response	Patient perception of improvement AND change on at least one outcome measure [high MCID threshold: I-RODS ≥ 8 centile points (or maximum I-RODS score), grip strength ≥ 14 kPa, MRC-SS ≥ 4 points (or maximum MRC-SS score)] or combined outcome measures [I-RODS ≥ 4 centile points and either MRC-SS ≥ 2 points or grip strength ≥ 8 kPa], assessed within 6 months of treatment initiation or change.
Optimal Response	Recovery as close as possible to the pre-CIDP functional status, excluding potential residuals, and fulfilling NEDA 2 criteria sustained for at least 6 months.
Relapse	Patient perception of worsening AND decline in disability AND impairment fulfilling IN-EDA definition following a period of stability or improvement, without a condition that better explains change in function and impairment, over a period of 12 weeks or less.
Possible Relapse	Patient perception of worsening AND decline in disability OR impairment following a period of stability or improvement over a period of 12 weeks or less.
Refractory	Absence of minimal response OR relapse leading to/requiring treatment discontinuation after adequate dosing and duration of the administered therapy.
Remission	No signs of disease activity (IN-NEDA 2) with or without residuals without immunosuppressive/immunomodulatory therapy for at least 12 months.
Residuals	Signs, symptoms, or impairments related to CIDP that persist despite adequate immunosuppressive/immunomodulatory therapies and are present in a patient who otherwise has no evidence of disease activity (IN-NEDA).

↘ DEFINITIONS FOR EVERYDAY OR NON-MEDICAL USE

The definitions in the table below are meant for informal use to help guide everyday conversations about CIDP. These simplified definitions do not replace the official definitions used by healthcare professionals, which reference validated clinical measurements to support each definition. Instead, they supplement those definitions and provide an easier way to talk about CIDP to non-healthcare professionals. These definitions should still be used as written without modification when possible.

Term	Simplified Definition
IN-EDA	This means that it is likely that your immune system is actively injuring your nerves. • You feel that your neuropathy is worse, and there is a measurable negative change in strength, function, or both. • You and your doctors should also make sure there isn't another reason for the change.
IN-NEDA 1	You do not feel your neuropathy symptoms worsening and for at least 6 months, you either: • Haven't gotten worse in your daily function as measured by your doctor, OR • Haven't gotten worse in your strength as measured by your doctor.
IN-NEDA 2	You do not feel your neuropathy symptoms worsening and for at least 6 months, you: • Haven't gotten worse in your daily function as measured by your doctor, AND • Haven't gotten worse in your strength as measured by your doctor.
Minimal Response	You feel that your neuropathy is a little better and there is a small improvement in strength or function as measured by your doctor. This happens within 3 months of starting treatment.
Partial Response	You feel your neuropathy is better and show a bigger improvement in at least one test of strength or function as measured by your doctor. This happens within 6 months.
Optimal Response	Your neuropathy has improved a lot—almost back to how you were before your symptoms initially began. You also meet IN-NEDA 2 criteria and stay that way for at least 6 months.
Relapse	After getting better or stable, your neuropathy symptoms come back. You feel worse and tests of function and strength from your doctor have gone down. This happens over 12 weeks or less.

Refractory	The treatment doesn't work at all—or it stops working and has to be stopped—despite giving it a full try.
Remission	Your neuropathy is quiet. You meet IN-NEDA 2 criteria (no signs of disease), and you've been off all immune treatments for at least 12 months. You may still have some residuals.
Residuals	These are leftover symptoms or problems with strength or function that stay even when the disease is no longer active. Residuals are due to old nerve injuries that are not expected to improve with immune drugs. Supportive interventions, like physical therapy, may be helpful.

↘ TERMS DEFINED IN 10 WORDS OR FEWER

These definitions are simplified (10 words or fewer) to support easy references in casual settings and help patients understand their CIDP journey. They do not replace or alter the official clinical definitions.

Term	Defined in 10 words or fewer
IN-EDA	You feel symptoms and tests show strength or function decline.
IN-NEDA 1	You do not feel worse; no worsening in strength or function.
IN-NEDA 2	You do not feel worse; no worsening in strength and function.
Minimal Response	You feel a bit better with small test improvements.
Partial Response	You feel better with bigger test improvements.
Optimal Response	Improved as much as possible (some residuals may persist); no disease signs for 6 months.
Relapse	Symptoms return and tests show you're getting worse.
Refractory	Treatment doesn't help or must be stopped.
Remission	No disease signs for 12+ months without treatment.
Residuals	Ongoing symptoms even without active disease.

↘ ABOUT

While we gathered CIDP patients from all corners of the world at the Leadership Collaborative, we were able to identify a common journey with CIDP that is reflected in the patient journey map on this page.

↘ SCAN HERE TO DOWNLOAD CIDP JOURNEY MAP