# CIDP in Focus
## Advancing Precision Across the Patient Journey from Early Recognition to Downstream Management

CLINICAL POINTS RESOURCE

*Supported through an independent educational grant from argenx.*

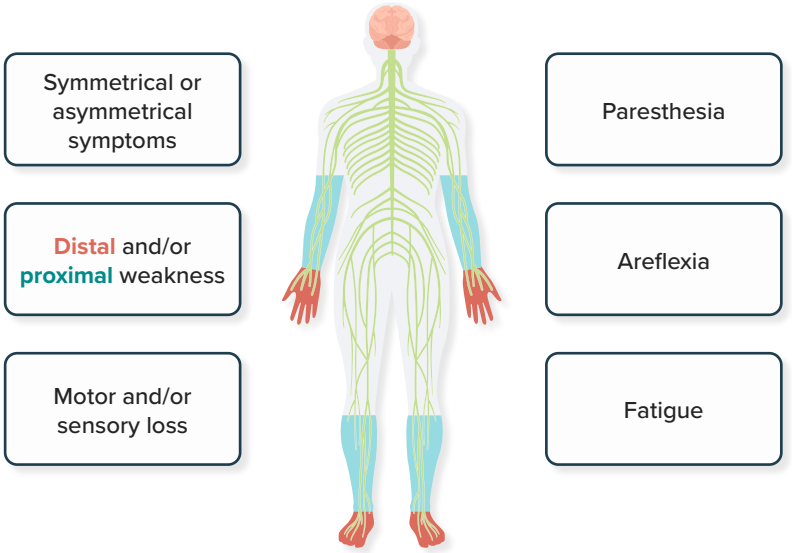


CME OUTFITTERS

This **resource guide** accompanies the **CIDP CaseWise™ Initiative by CME Outfitters**, which addresses key aspects of CIDP, including disease prevalence, pathophysiology, diagnosis, treatment strategies, emerging therapies, and patient-centered care. The actionable resources linked below are intended to help improve quality of care and outcomes for patients with CIDP.

| Action Item | Scope of the CIDP Problem |
|---|---|
| Evaluate CIDP **prevalence, presentation, and progression** | |

### Presentation of CIDP

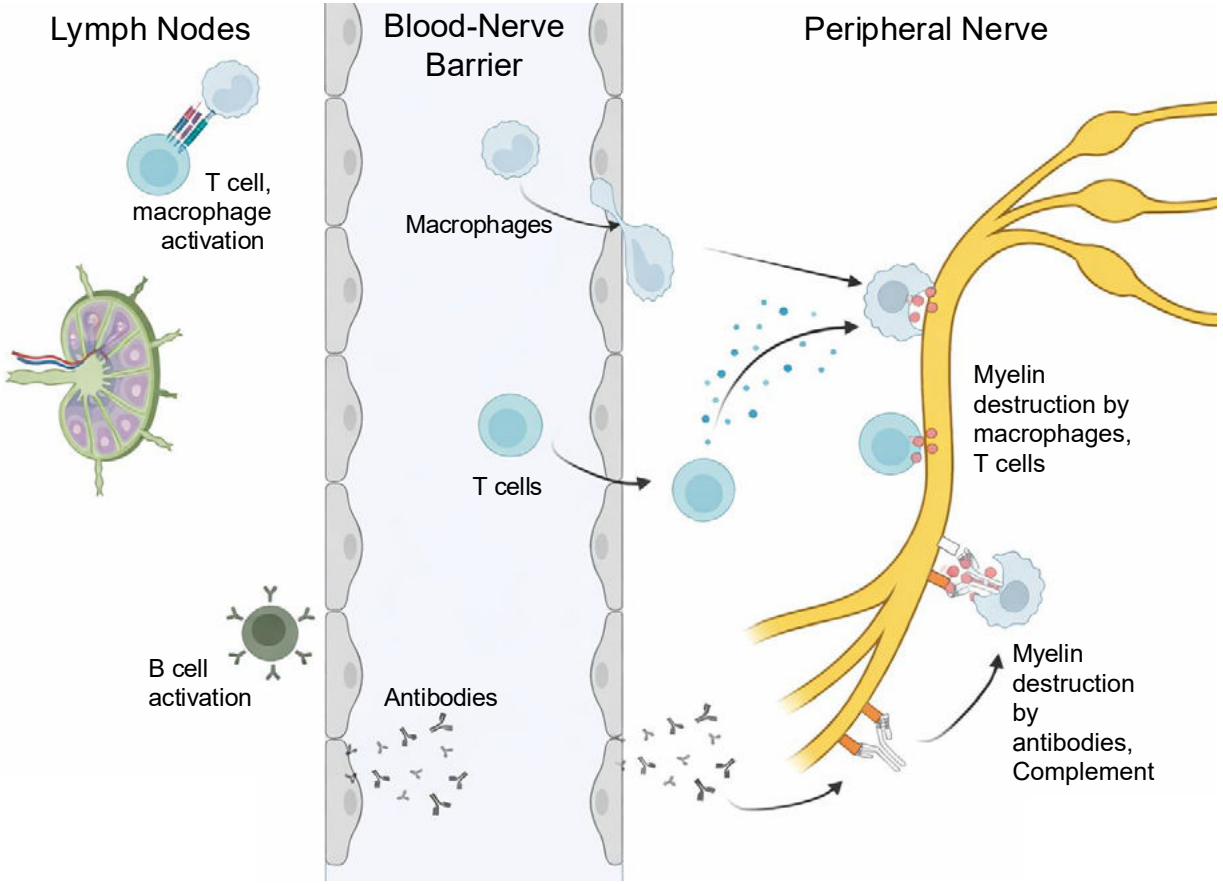



- CIDP is a rare disease: 0.8-8.9 cases per 100,000
- Three categories of progression
  - **Monophasic:** single bout of CIDP for several months to years with spontaneous recovery
  - **Chronic relapsing:** symptoms continue to progress or worsen with partial spontaneous improvement or remissions between relapses
  - **Chronic progressive:** constant worsening without improvement
- After flare-ups, patients often left with discomfort, weakness, residual numbness even with complete remission
- Leads to substantial physical, psychosocial, and economic burden

**CIDP** = chronic inflammatory demyelinating polyneuropathy.

Broers MC, et al. *Neuroepidemiology*. 2019;52(3-4):161-172. Brun S, et al. *Immuno*. 2022;2(1):118-131. Querol L, et al. *J Neurol*. 2021;268(10):3706-3716.

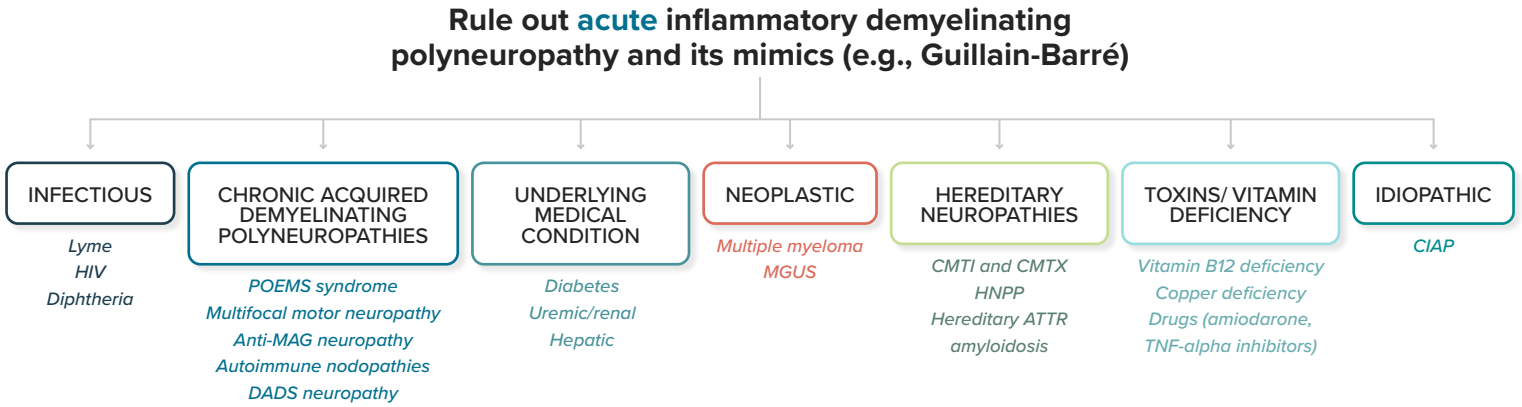
DOWNLOADABLE RESOURCE: **Incidence, Prevalence, and Treatment Patterns in CIDP: Data Analysis of U.S. Claims**

| Action Item | Scope of the CIDP Problem |
|---|---|
| **Assess the presumed mechanism that leads to CIDP** (immune system attacks myelin sheaths in peripheral nerves) |  |



Kohle F, et al. *Ther Adv Neurolog Disord*. 2023;16:1756286422137129.

DOWNLOADABLE RESOURCE: **A Pathophysiological and Mechanistic Review of CIDP Therapy.**

| Action Item | Differential Diagnosis |
|---|---|
| Rule out acute inflammatory demyelinating polyneuropathy and its mimics |  |



**ATTR** = amyloid transthyretin; **CIAP** = chronic idiopathic axonal polyneuropathy; **CMTI** = Charcot-Marie-Tooth Type 1; **CMTX** = Charcot-Marie-Tooth X-linked; **HCPP** = hereditary neuropathy with liability to pressure palsies; **HIV** = human immunodeficiency virus; **MAG** = myelin-associated glycoprotein; **MGUS** = monoclonal gammopathy of undetermined significance; **POEMS** = polyneuropathy, organomegaly, endocrinopathy, monoclonal plasma cell disorder, skin changes.

Eftimov F, et al. *Brain*. 2020;143(11):3214-3224. Van den Berg PYK, et al. *J Periph Nerve Syst*. 2021;26:242-268.

DOWNLOADABLE RESOURCE: **Diagnosis of Chronic Inflammatory Demyelinating Polyneuropathy**

| Action Item | Clinical Criteria |
|---|---|
| Consider **typical CIDP and variants** | |

## Typical



- Muscle weakness
  - *Symmetric*
  - *Proximal AND distal*
  - *Upper AND lower extremities*
- Sensory involvement (≥ 2 limbs)
- Progressive or relapsing
- Absent or reduced **reflexes** in all limbs
- Developed over ≥ **8 weeks**

## Variants

- *Progressive or relapsing*
- *Developed over ≥8 weeks*
- *Reflexes may be normal in unaffected limb*

| | | |
|---|---|---|
|  | **MOTOR** | *No sensory involvement, proximal and distal, symmetric in 4 limbs* |
|  | **SENSORY** | *No motor involvement, symmetric in 4 limbs* |
|  | **FOCAL** | *Sensorimotor involvement in only 1 limb* |
|  | **MULTI-FOCAL** | *Sensorimotor, asymmetric, upper limb predominance, ≥2 limbs* |
|  | **DISTAL** | *Sensorimotor, lower limb predominance, ≥2 limbs* |

Van den Bergh PYK, et al. *J Peripher Nerv Syst*. 2021;26(3):242-268.

DOWNLOADABLE RESOURCE: **Defining "Typical" and "Atypical" Variants of CIDP (Video)**

| Action Item | Identifying Clinical Red Flags |
|---|---|
| **Look for red flags** pursuant to EAN/PNS diagnostic criteria | **TO AVOID UNDERDIAGNOSIS**<br>• Consider CIDP on the differential for patients with proximal muscle weakness<br>• Duration of symptoms helps differentiate CIDP from Guillain-Barré syndrome (GBS)<br>» *Progression 8 or more weeks after symptom onset strongly indicates CIDP over GBS*<br><br>**TO AVOID OVERDIAGNOSIS**<br>*Look for clinical red flags*<br>• Pain predominance<br>• Autonomic features<br>• Dyspnea<br>• Cranial nerve involvement<br>• Systemic features<br>• Family history<br>• Subacute, low-frequency tremor, marked ataxia, distal predominance<br>• Lean on EAN/PNS criteria |

DOWNLOADABLE RESOURCE: **EAN/PNS Guideline on Diagnosis and Treatment of CIDP.**

| Action Item | Electro-Diagnostic Criteria |
|---|---|

**Be sure to have the 2021 EAN/PNS Criteria** beside you when conducting the electrodiagnostic study

| MOTOR ABNORMALITIES | |
|---|---|
| Motor distal latency prolongation | Absence of F-waves |
| Motor conduction block | Distal CMAP prolongation |
| Conduction slowing | F-wave latency prolongation |
| Temporal dispersion | |

| SENSORY ABNORMALITIES |
|---|
| Prolonged distal latency |
| Reduced SNAP amplitude |
| Slowed conduction velocity |

**CIDP:** Clinical criteria + motor criteria ≥ 2 nerves + sensory abnormalities in ≥ 2 nerves, or possible CIDP + ≥ supportive criteria

**CMAP** = compound muscle action potential; **EAN/PNS** = European Academy of Neurology/Peripheral Nerve Society; **SNAP** = sensory nerve action potential.

DOWNLOADABLE RESOURCE: **EAN/PNS Guideline on Diagnosis and Treatment of CIDP.**

| Action Item | Electro-Diagnostic Pitfalls |
|---|---|
| **Strictly adhere to 2021 EAN/PNS electro-diagnostic criteria** to avoid diagnostic pitfalls | |

| PITFALLS[1-4] | SOLUTIONS |
|---|---|
| Loose interpretation of demyelinating criteria | *Strictly adhere to 2021 EAN/PNS electrodiagnostic criteria*[5] |
| Misinterpretation of NCS | **CAUTION FOR FOLLOWING NCS FINDINGS[1-3]**: Amplitude-dependent CV slowing; Amplitude-independent CV slowing in patients with diabetes; CV slowing limited to compressible / entrapment sites; Absence of conduction block |
| Suboptimal technique when performing NCS | *Warm the patient to a **minimum of 33°C**, confirm **supramaximal stimulation**, **minimize co-stimulation** of adjacent nerves*[6] |

**CV** = conduction velocity; **NCS** = nerve conduction studies.

**1.** Allen JA. *Neurol Ther*. 2020;9(1):43-54. **2.** Allen JA, et al. *Muscle Nerve*. 2018;57(4):542-549. **3.** Allen JA, et al. *Neurology*. 2015;85(6):498-504.
**4.** Stino AM, et al. *Muscle Nerve*. 2021;63(2):157-169. **5.** Van den Bergh PYK, et al. *J Peripher Nerv Syst*. 2021;26(3):242-268.
**6.** Koo YS, et al. *J Clin Neurol*. 2012;8(1):1-14.

DOWNLOADABLE RESOURCE: **EAN/PNS Guideline on Diagnosis and Treatment of CIDP.**

| Action Item | Supportive Studies | | | |
|---|---|---|---|---|
| **Use supportive criteria** for patients with possible CIDP **[need at least 2]** | **IMAGING**<br>• MRI can add value in assessing proximal nerves and the brachial and lumbosacral plexuses<br>• Ultrasound can evaluate focal areas of cross-sectional area enlargement | **CSF**<br>• Albuminocytologic dissociation in CSF (normal leukocytes, < 10 cells/µL, with elevated protein) is often seen in CIDP<br>• For individuals age > 50 years, consider values up to 0.6 g/L as acceptable | **NERVE BIOPSY**<br>• Rarely needed<br>• May help in complex cases by identifying demyelinating features and evidence of inflammation | **SEROLOGY TESTING**<br>*(not supportive for diagnosis)*<br>• Serum monoclonal proteins using serum protein electrophoresis with immunofixation should be performed in every individual with suspected CIDP<br>• Anti-MAG (myelin-associated glycoprotein) antibodies should be tested in distal CIDP |

***Note:** Response to treatment can also serve as supportive criteria*

- Improvement after IVIg, PLEX, or steroids
- Needs improvement on at least 1 disability scale and 1 impairment scale
- Patient-reported improvement alone does not count

**CSF** = cerebrospinal fluid; **IVIg** = intravenous immunoglobulin; **MRI** = magnetic resonance imaging; **PLEX** = plasma exchange.

DOWNLOADABLE RESOURCE: **EAN/PNS Guideline on Diagnosis and Treatment of CIDP.**

| Action Item | CSF Analysis |
|---|---|
| **Avoid** overreliance on mild/moderate albuminocytologic dissociation, especially with normal NCS<br><br>**Consider** alternative explanations for elevated CSF protein (e.g., age, diabetes, spondylotic diseases, hereditary neuropathies) | • **Albuminocytologic dissociation** is common<br>  ◦ Occurs in up to 75% of patients with CIDP[1-4]<br>  ◦ Occurs in up to 50% of misdiagnosed cases of CIDP (generally mild)[2]<br>• Limitation of CSF protein levels in CIDP<br>  ◦ **Unknown specificity:** cannot distinguish between CIDP and its many mimics<br>  ◦ **Lack of well-established cutoffs** for older patients (age > 50 years)<br>  ◦ Elevated CSF protein must be interpreted with caution for **patients with diabetes or spinal stenosis** |

***Remember:***

- Remember: Higher normal CSF reference values may improve specificity:
  - » Age ≤ 50 years: < 50 mg/dL
  - » Age > 50 years: < 60 mg/dL

You do not need CSF if patient meets clinical and electrodiagnostic criteria

**1.** Van den Bergh PYK, et al. *J Peripher Nerv Syst*. 2021;26(3):242-268. **2.** Allen JA, et al. *Neurology*. 2015;85(6):498-504.
**3.** Allen JA. *Neurol Ther*. 2020;9(1):43-54. **4.** Kieseier BC, et al. *Nat Rev Dis Primers*. 2018;4(1):31.

DOWNLOADABLE RESOURCE: **CSF Diagnostics: A Potentially Valuable Tool in Neurodegenerative and Inflammatory Disorders Involving Motor Neurons: A Review**

| Action Item | First-Line Therapies |
|---|---|
| For motor CIDP, **consider IVIg** for first-line therapy (may worsen with steroids)<br>**Remember:** ~10-15% of patients remain refractory to first-line therapies | **FIRST-LINE THERAPIES**<br>**INITIAL**<br>• IVIg<br>• Plasma exchange (PLEX)<br>• Corticosteroids<br>**MAINTENANCE**<br>• IVIg<br>• Subcutaneous Ig (SCIg)<br>• Facilitated subcutaneous Ig (fSCIg)<br>• Glucocorticoids<br>**LIMITATIONS**<br>**GENERAL:** Approximately 10–15% of patients remain refractory to first-line therapies<br>**SPECIFIC**<br>• Corticosteroids – side effects and safety concerns *(e.g., immunosuppression, diabetes, osteoporosis)*<br>• IVIg – wearing-off effects<br>• PLEX and IVIg – Logistical and adherence challenges *(i.e., due to travel to infusion centers)* |

Van den Bergh PYK, et al. *J Peripher Nerv Syst*. 2021;26(3):242-268.

DOWNLOADABLE RESOURCE: **EAN/PNS Guideline on Diagnosis and Treatment of CIDP.**

| Action Item | Objective Assessments |
|---|---|
| **Improve the effective and appropriate utilization of treatment** by using objective indicators | **CIDP ASSESSMENTS** |

**DISABILITY**

- **INCAT:** Inflammatory Neuropathy Cause and Treatment disability scale
- **I-RODS:** Inflammatory Rasch-Built Overall Disability Scale

**STRENGTH**

- **Grip strength**
- **Manual muscle testing:** use Medical Research Council (MRC) grading system

**GAIT**

- **TUG:** Timed Up and Go test
- **10MWT:** 10-meter walk test
- **6MWT:** 6-minute walk test
- **Wearable sensors**

**QoL***

- **CAPPRI:** Chronic Acquired Polyneuropathy Patient-Reported Index

**Does not include a complete and exhaustive list of QoL assessments*

**QoL** = quality of life.

Allen JA, et al. *JAMA Neurol*. 2020;77(9):1159-1166.

DOWNLOADABLE RESOURCES:

**Getting Closer to Patients: the INCAT Overall Disability Sum Score**

**IRODS for Immune-Mediated Peripheral Neuropathies**

**Evaluating Grasp Function in Patients with CIDP Using Dynaamometers**

| Action Item | Conventional vs Facilitated SCIg |
|---|---|

**Consider facilitated SCIg** as an alternative to IVIg when appropriate

**BACKGROUND**

- Hyaluronidase increases SQ permeability to IgG
- Higher concentrations of IgG (shorter / less volume)

**STUDY DESIGN**

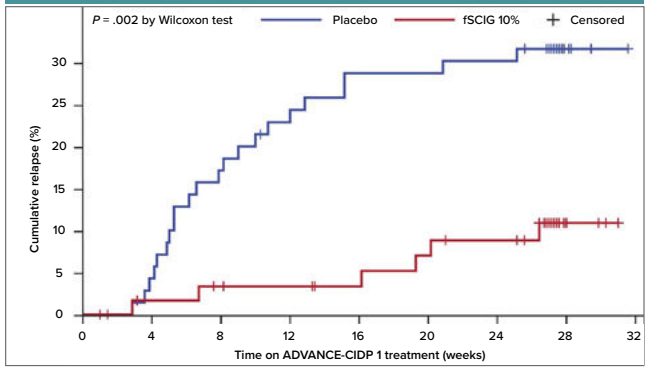
- Adults with probable or definite CIDP (N = 132; INCAT scores 0 to 7) received IVIg ≥ 12 weeks before being randomized 1:1 to fSCIg (every 2–4 weeks) or placebo

**RESULTS**

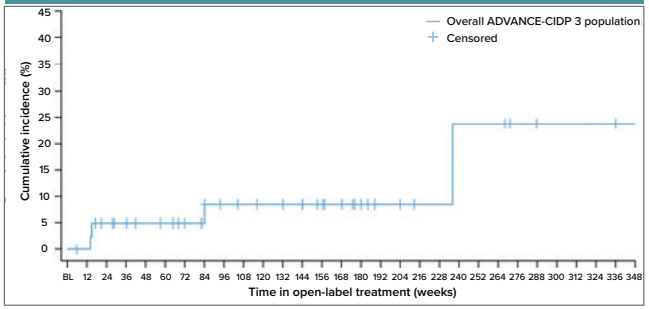
- Double blind: 21.8% reduction in relapse rate (*P* = .0045)
- Open label extension: 13% relapsed during follow-up; annual relapse rate ~4.5%

**COMMON SIDE EFFECTS:** Injection site reactions, headache, nausea, fatigue

**PERCENTAGE RELAPSE fSCIg PLACEBO**




**LONG TERM PROBABILITY OF RELAPSE**

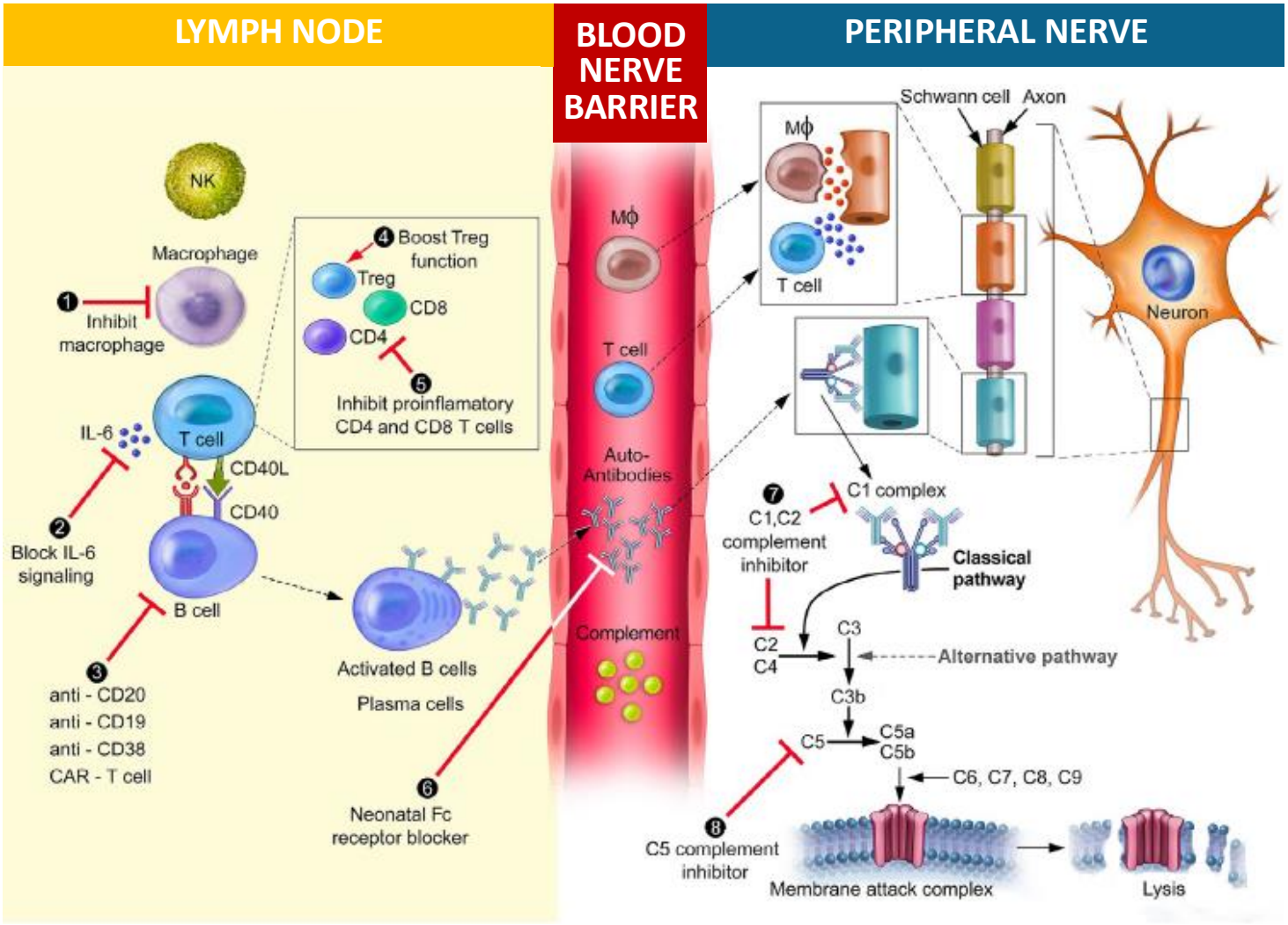



**fSCIg** = hyaluronidase-facilitated subcutaneous immunoglobulin.

DOWNLOADABLE RESOURCE: **Fundamentals of subcutaneous immunoglobulin therapy**

| Action Item | New and Emerging Therapeutic Targets in CIDP |
|---|---|

**Recognize the therapeutic targets** of new and emerging therapeutic agents for CIDP




Gable KL, Li Y. *Muscle Nerve*.2025;72(2):201-211.

VIDEO RESOURCE: **Dr. Karissa Gable takes you through this graphic step-by-step.**

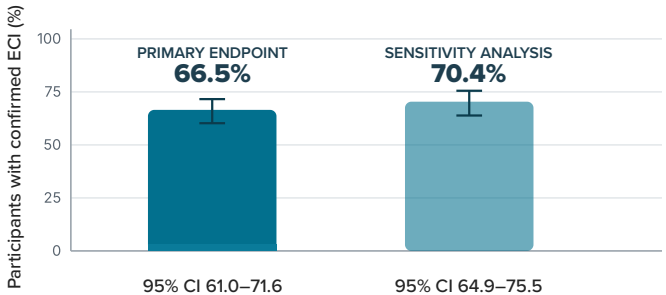
## Action Item

**Evaluate results** from the ADHERE trial that led to FDA approval of **efgartigimod**, and the long-term extension data that followed

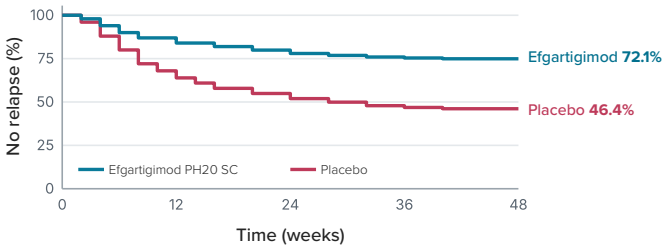
## Approved FcRn Inhibitor

### ADHERE TRIAL (efgartigimod)

#### STAGE A (OPEN LABEL)
Confirmed ECI




#### STAGE B (RANDOMIZED WITHDRAWAL)
Time to First aINCAT Deterioration




**61% reduced risk of relapse** — 27.9% relapsed with efgartigimod vs 53.6% with placebo (HR = 0.39; *P* = .0001)

**Common TEAEs (mostly mild to moderate):** injection site reactions, respiratory infections, headache

### Long-Term Follow-Up (ADHERE+)

- Open-label extension of ADHERE; 229 enrolled, 228 treated. Mean time on study ~61 weeks; maximum combined exposure 187 weeks across ADHERE and ADHERE+.
- Improvement continued over time — mean change from run-in baseline to week 36: **−1.2 points aINCAT, +8.8 centiles I-RODS, +17.5 kPa grip strength**.
- 34.7% (68/196) reached an aINCAT score of ≤1 at some point during the extension.
- TEAE incidence did not increase with prolonged exposure; serious TEAEs 15.4%, discontinuation due to TEAE 7.9%.

### Administration Options

- Prefilled syringe (1,000 mg/10,000 units) approved April 2025 for **weekly self- or caregiver administration** over ~20–30 seconds, following training in subcutaneous injection technique.
- Vial formulation (1,008 mg/11,200 units) administered weekly by a health care professional over ~30–90 seconds.
- Observe patients for hypersensitivity reactions for at least 30 minutes after administration.
- A missed dose may be given up to 3 days after the scheduled time; otherwise resume the original schedule.

**aINCAT** = adjusted Inflammatory Neuropathy Cause and Treatment disability score; **CI** = confidence interval; **ECI** = evidence of clinical improvement; **HR** = hazard ratio; **I-RODS** = Inflammatory Rasch-built Overall Disability Scale; **TEAE** = treatment-emergent adverse event.

Allen J, et al. *J Peripher Nerv Syst*. 2024;29:S181. Allen JA, et al. *Lancet Neurol*. 2024;23(10):1013-1024. Allen JA, et al. *J Peripher Nerv Syst*. 2026;31(3):e70140. Efgartigimod alfa and hyaluronidase-qvfc injection, for subcutaneous use [package insert]. U.S. Food and Drug Administration Website. https://www.accessdata.fda.gov/drugsatfda_docs/label/2026/761304s018lbl.pdf. Revised May 2026.

DOWNLOADABLE RESOURCE: **A Plain Language Summary of the ADHERE trial (for Shared Decision-Making with Patients)**

| Action Item | Patient Resources |
|---|---|
| Consider questions to ask your clinician at your next appointment | The **GBS \| CIDP Foundation International** is a global nonprofit organization supporting individuals and their families affected by GBS, CIDP, multifocal motor neuropathy (MMN), and related conditions through a commitment to support, education, research, and advocacy.<br><br>GBS \| CIDP Foundation International™<br><br>DOWNLOADABLE RESOURCE: **Questions to Ask Your Healthcare Provider** |
| **Review this whiteboard animation** to get a better sense of what causes CIDP | **Chronic Inflammatory Demyelinating Polyneuropathy 101**<br><br><br><br>Source: GBS-CIDP Foundation International |

| Action Item | Patient Resources |
| --- | --- |
| **Learn how other people with CIDP are taking steps to improve quality of life** by viewing the 2026 series, “Rare, Well Done” (subscription to Peacock TV required) | **Rare, Well Done** is a 2026 reality series streaming on Peacock TV that focuses on people living with CIDP. The show features the “Thrive Team” of experts helping patients regain joy and independence. (Subscription required)   |