Overview and Treatment of Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)
Description of CIDP

- CIDP is a form of peripheral neuropathy, characterized by demyelination with secondary axonal dysfunction and consequential failure of nerve conduction in motor and sensory nerves.
- CIDP is immune-mediated and progressive, but if caught early, i.e. before axonal damage, can be reversed by medical treatment, e.g., immunoglobulin (Ig) or corticosteroid therapy or plasma exchange.\(^1\) If allowed to progress, permanent axonal damage can result.

CIDP: chronic inflammatory demyelinating polyneuropathy, Ig: immunoglobulin

Prevalence of CIDP

- Peak prevalence is between 40 to 60 years of age\(^1\) with rates ranging from 1.0 to 8.9 per 100,000 in different regions.\(^2-6\)
- It is less common in children, with prevalence estimated to be 0.5 per 100,000.\(^7\)
  - Typically children have a relapsing-remitting course and have a more favorable prognosis than adults.

CIDP: chronic inflammatory demyelinating polyneuropathy

Symptoms of most common form of CIDP

- CIDP appears as symmetrical weakness in both proximal and distal muscles, which progressively worsens for >2 months.¹
- Usually associated with impaired sensation, thus further reducing the ability to control and engage muscles of the legs, feet, arms and hands.
- Patients may be unable to lift themselves from a chair, maintain balance, or handle small and delicate items.

CIDP: chronic inflammatory demyelinating polyneuropathy
Prognosis of CIDP

- CIDP is slowly progressive in the majority of patients and is relapsing-remitting in others.¹

- Early diagnosis and treatment is vital to prevent irreversible axonal loss and improve functional recovery and quality of life.²

- CIDP that is not treated leads to accumulating disability that requires:
  - Physical therapy
  - Orthotic devices
  - Long-term treatment

- If left untreated, 30% of patients will progress to wheelchair dependence.³

CIDP: chronic inflammatory demyelinating polyneuropathy

Diagnostic guidelines on CIDP

- The joint task force of the European Federation of Neurological Societies (EFNS) and Peripheral Nerve Society (PNS) has developed guidelines on CIDP.¹

Diagnosis
- Clinical
- Electrophysiology (evidence for demyelination)
- CSF analysis
- Nerve biopsy
- Supportive evidence
  - MRI
  - Response to immunomodulatory treatment

CIDP: chronic inflammatory demyelinating polyneuropathy, CSF: cerebrospinal fluid, MRI: Magnetic resonance imaging

CIDP - differential diagnoses

• Diabetic neuropathy (DN)
  – Some patients with diabetes mellitus may have additional CIDP and respond to IVIG
  – May need to watch out for progression and early motor symptoms in DN in order not to miss CIDP

• Late-onset hereditary neuropathy
  – Watch out for family history, foot and spine deformities, history of poor sports performance in childhood
Treatment guidelines on CIDP

Treatment¹

- Clinical trials demonstrate therapy effectiveness

---

CIDP: chronic inflammatory demyelinating polyneuropathy, IVIG: intravenous immunoglobulin
Treatment options for CIDP

- In 2010, immunoglobulin (IgG) therapy was the most common treatment used for treatment of CIDP in the United States\(^1\)

- Immunoglobulins are a common treatment for CIDP:
  - Easy to use
  - Effectiveness shown in clinical trials

CIDP: chronic inflammatory demyelinating polyneuropathy, IgG: immunoglobulin G

Immunoglobulin therapy in CIDP

- Induction and maintenance doses assessed in several clinical trials and used in clinical practice

Induction dose: 2 g/kg over 2-5 days
Maintenance dose: 1 g/kg approximately every 3 weeks

- Guideline recommendation:
  - When stable on IVIG, a patient’s dose should be reduced to the lowest dose that continues to provide clinical benefit

- Treatment response measured by change in:
  - Inflammatory Neuropathy Cause and Treatment (INCAT) scale
  - MRC sum score
  - RODS
  - Functional assessments such as grip strength and walking tests

CIDP: chronic inflammatory demyelinating polyneuropathy, INCAT: inflammatory neuropathy cause and treatment, IVIG: intravenous immunoglobulin, MRC: Medical Research Council

Additional thoughts on treating CIDP

- Rapid diagnosis is essential to limit axonal dysfunction and demyelination
  - Identification of biological markers may aid diagnosis
- Understanding disease processes may help optimize and develop new treatments
- Alternative routes of IgG administration are under investigation
- Investigations to understand more about CIDP and its treatment are continuing