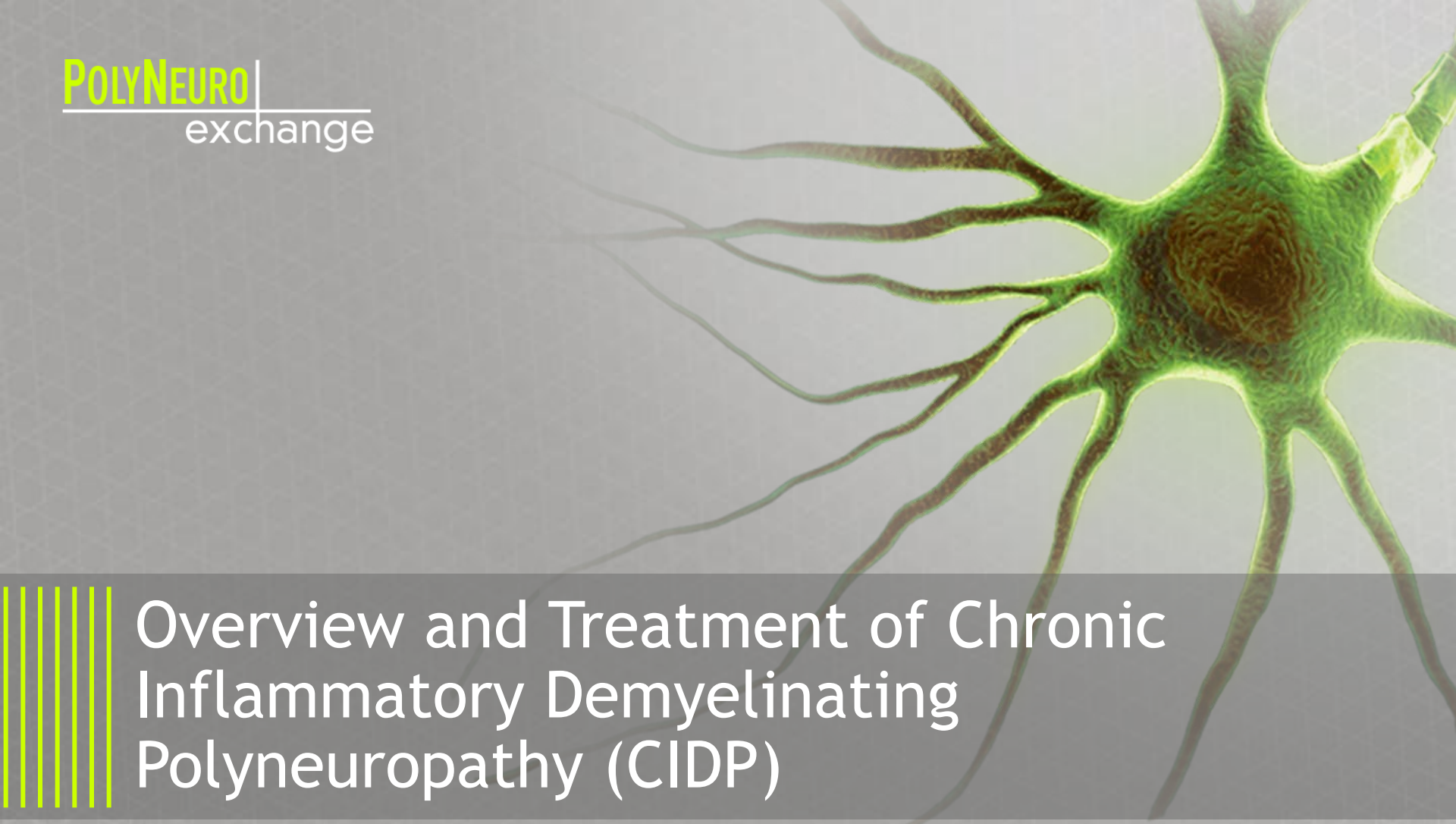
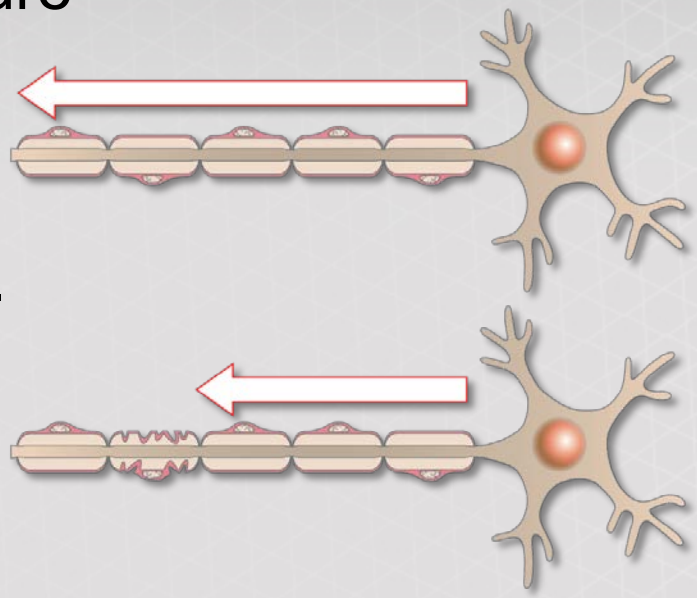
PolyNeuro exchange



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

1. Yoon M-S, *et al.* Ther Adv Neurol Disord 2011;4:193-200

Prevalence of CIDP

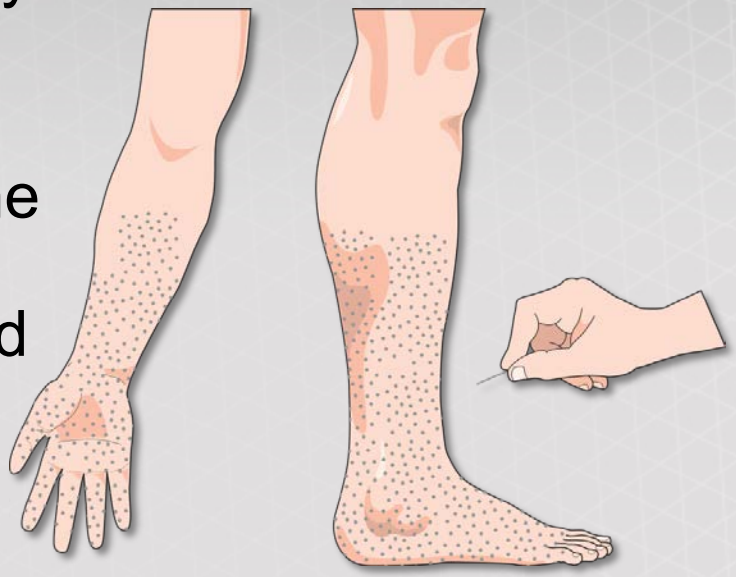
- Peak prevalence is between 40 to 60 years of age[1] with rates ranging from 1.0 to 8.9 per 100,000 in different regions.[2-6]
- It is less common in children, with prevalence estimated to be 0.5 per 100,000.[7]
  - Typically children have a relapsing-remitting course and have a more favorable prognosis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

CIDP: chronic inflammatory demyelinating polyneuropathy

1. Robertson EE, Donofrio PD. Curr Treat Options Neurol 2010;12:84-94
2. Lunn MP, *et al*. J Neurol Neurosurg Psychiatry 1999;66:677-680
3. McLeod JG, *et al.* Ann Neurol 1999;46:910-913
4. Mygland A, Monstad P. Eur J Neurol 2001;8:157-165
5. Iijima M, *et al*. J Neurol Neurosurg Psychiatry 2008;79:1040-1043
6. Laughlin RS, *et al*. Neurology 2009;73:39-45
7. Markowitz JA, *et al*. Neurology 2008;71: e74–e78

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

1. Yoon M-S, *et al.* Ther Adv Neurol Disord 2011;4:193-200

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

1. Lewis R. *http://emedicine.medscape.com/article/1172965-* Accessed January 2012
2. Robertson EE, Donofrio PD. Curr Treat Options Neurol 2010;12:84-94
3. GBS-CIDP Foundation. http://www.gbs-cidp.org/home/cidp/cidp/ Accessed February 2012

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

Mandatory

Not Mandatory

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

1. Van den Bergh PYK, *et al.* Eur J Neurol 2010;17:356-363

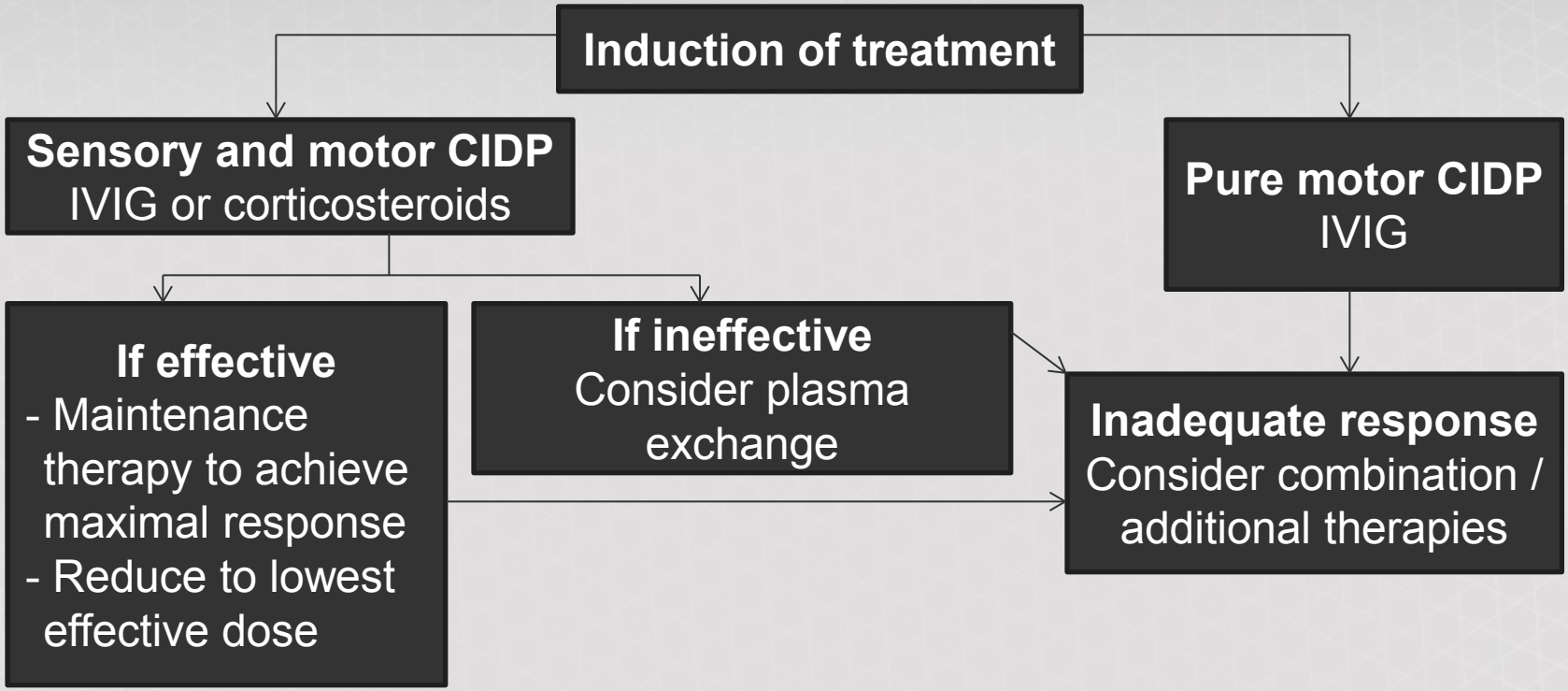
CIDP – differential diagnoses

- 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

Treatment[1]

- Clinical trials demonstrate therapy effectiveness

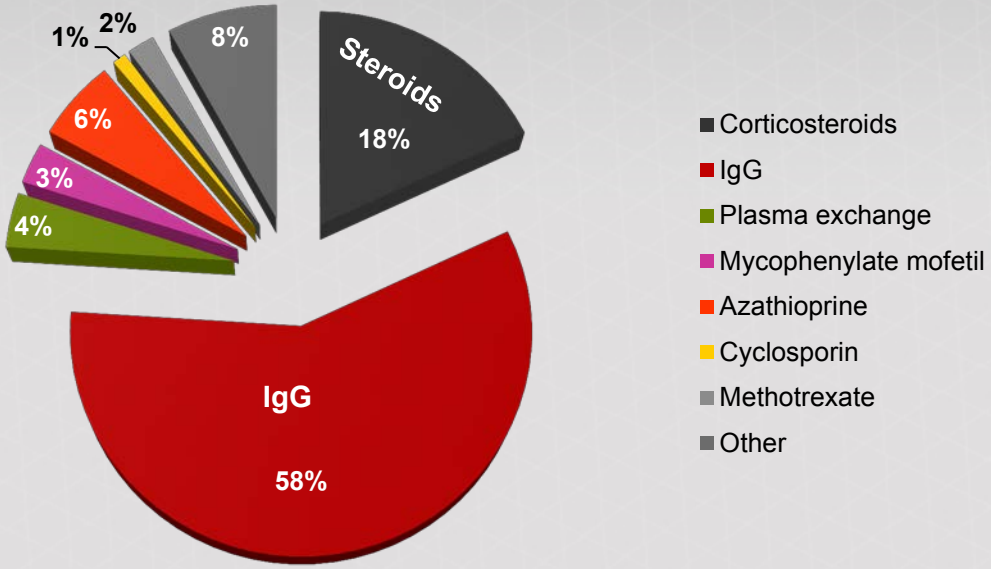


CIDP: chronic inflammatory demyelinating polyneuropathy, IVIG: intravenous immunoglobulin

1. Van den Bergh PYK, *et al.* Eur J Neurol 2010;17:356-363

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

1. Koski CL, *et al.* 2010. 2010 survey of patients with chronic inflammatory demyelinating polyneuropathy in the USA: Poster presented at AAN 2011

Immunoglobulin therapy in CIDP

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

- Guideline recommendation:
  - When stable on IVIG, a patient's dose should be reduced to the lowest dose that continues to provide clinical benefit[1]
- 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

1. Van den Bergh PYK, *et al.* Eur J Neurol 2010;17:356-363

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

CIDP: chronic inflammatory demyelinating polyneuropathy, IgG: immunoglobulin G, IVIG: intravenous immunoglobulin, SCIG: subcutaneous immunoglobulin