Overview and Treatment of Guillain-Barré Syndrome (GBS)
Description of GBS

- GBS is an acute, immune-mediated polyneuropathy.
- GSB is the most common cause of acute, non-traumatic neuromuscular paralysis worldwide, with an estimated annual incidence of about 2 per 100,000 people.¹⁻³
- GBS affects all ages, ethnicities and genders.
- GBS is more common in men and the elderly.³

GBS: Guillain-Barré Syndrome

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General features
• Acute onset with maximal disability reached within 4 weeks of onset for 90% of patients.¹
• Majority of patients report an antecedent event 28 days before onset; commonly upper respiratory tract infection or Campylobacter jejuni infection.¹⁻³

Symptoms, signs and tests
• Bilateral and symmetrical loss of sensation and weakness which usually begins in feet and spreads upward and may lead to difficulty in breathing.
• Reflexes are lost.
• Spinal fluid protein is frequently elevated.
• Nerve conduction studies show a neuropathy.
Prognosis of GBS

- GBS is a monophasic, self-limiting disease, that worsens over 2-4 weeks and then starts to improve.
- Strong evidence shows that treatment with either IVIg or Plasma exchange (PE) improves recovery and outcome\(^1\).
- Recovery period varies and may take weeks to years.
- Recovery is affected by age, antecedent gastroenteritis, speed of onset, maximal disability and nerve conduction evidence of axon loss.\(^2,3\)
- Erasmus GBS Outcome Scale can be used to select GBS patients at risk for a poor prognosis.\(^2\)
- Approximately 5% of GBS patients die and up to 20% have persistent disability, despite immunotherapy.\(^4\)

## Diagnosis of GBS

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<tr>
<th>Diagnostic criteria(^1,2)</th>
<th>Manifestation</th>
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| **Clinical features essential for diagnosis** | • Progressive weakness in arms and legs  
• Areflexia  
• Maximum weakness reached within 4 weeks |

| **Clinical features supporting diagnosis** | • Relatively symmetrical, progressive over days to four weeks  
• Autonomic features  
• Cranial nerve involvement  
• Absence of fever |

| **Clinical features not supporting diagnosis** | • Significant asymmetry  
• Early and persistent bowel or bladder disturbance |

| **Differential diagnoses** | • Toxic neuropathies  
• Hereditary neuropathies (e.g. porphyria)  
• Inflammatory diseases (e.g. collagen vascular disease, Lyme disease)  
• Metabolic neuropathies (e.g. diabetes mellitus)  
• Carcinomatous meningitis  
• Other such as transverse myelitis, myasthenic crisis, acute rhabdomyolysis |

| **CSF and electrodiagnostic findings** | • Elevated protein with normal cell count (albumino-cytologic dissociation)  
• Electrodiagnostic studies showing a neuropathy |

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CSF: cerebrospinal fluid  
Treatment options for GBS

• Plasma exchange (PE) and IVIG are effective in the treatment of GBS.\(^1,2\)
  – PE: usually administered as one plasma volume, 50 mL/kg on five separate occasions over 1-2 weeks.
  – IVIG: total dose of 2g/kg delivered over 5 days or 0.4 g/kg/day x 5 days.

• Attentive and supportive care reduce morbidity and mortality.\(^1,3\)
  – Mechanical ventilation for respiratory paralysis.
  – Monitoring for hypertension, postural hypotension and cardiac arrhythmia.
  – Opioid analogs / other drugs for pain and sensory symptoms.
  – Subcutaneous heparin and support stockings for DVT risk.

• Emotional support and rehabilitation.

DVT: deep vein thrombosis, GBS: Guillain-Barré Syndrome, IVIG: intravenous immunoglobulin
2. Van der Meché FG, Schmitz PI. NEJM 1992;326(17):1123-1129
Outcome measures for GBS

- Outcome measures typically assess the following functions:

**Motor:**
- GBS disability score
- MRC sum score
- FIM Motor Scale
- Hand function tests
- Walking tests

**Sensory:**
- Visual Analog Pain Scale
- McGill Pain Questionnaire-Short Form
- Romberg’s test
- Pinprick, light touch and vibration perception tests

**Duration:**
- Length of ICU or hospital stay
- Duration of mechanical ventilation (if applicable)

**Other:**
- IgG serum levels
- Erasmus GBS Outcome Scale

International Guillain-Barré Syndrome Outcome Study (IGOS)\textsuperscript{1}

- IGOS is a worldwide prospective study being conducted by the Inflammatory Neuropathy Consortium of the Peripheral Nerve Society.
- Aim: To define biomarkers for disease activity and recovery and to develop prognostic models to predict the clinical course and outcome in individual patients with GBS.
- Study design: A prospective, observational multicenter study, including at least 1000 patients, with a follow-up period of at least one year.
- Expected results: a standardized clinical database and biobank with up to 3-year patient follow-up.
- These data will be available for researchers to determine processes of disease progression and recovery in GBS, to develop prognostic models, conduct selective therapeutic trials, and personalize treatment.

GBS: Guillain-Barré Syndrome, IGOS: International Guillain-Barré Syndrome Outcome Study

1. International GBS Outcome Study. \url{https://www.gbsstudies.org/about-igos} Accessed February 2013
Additional thoughts on GBS

• There is still a need for improved treatment options in GBS:
  – Up to 5% of patients with GBS die and ~15% are unable to walk after one year.

• A study of a second IVIG infusion in those with a poor prognosis is underway.
  – Anecdotal reports suggest that in some patients a 2nd IVIg dose may be beneficial.
  – One study suggests that patients with higher increments of IgG levels have better outcomes.\(^1\)

• Research on supplementary therapies that might protect the axon and/or promote the axonal regeneration are underway.