# Guillain–Barré syndrome

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## 1 Guillain–Barré syndrome (GBS) is the most frequent cause of subacute neuromuscular weakness in North America

The median incidence of GBS is about 1 per 100 000 person-years, with higher rates in older people (a 20% increase in the average GBS rate for every 10-year increase in age) and males.[1] Infections can trigger GBS (e.g., including those caused by *Campylobacter jejuni* and influenza virus). However, a large retrospective study published in 2013 found no increased risk of GBS after vaccination.[2]

## 2 Guillain–Barré syndrome usually presents with symmetric ascending weakness and hyporeflexia

Weakness usually begins in the lower extremities.[3] In primary care, patients most commonly present with acroparesthesias followed by subacute (< 4 wk), symmetric, flaccid weakness with hyporeflexia (Appendix 1, available at www.cmaj.ca/lookup/doi/10.1503/cmaj.202710/tab-related-content). Referral to the emergency department with consideration of inpatient admission to monitor for progressive weakness, respiratory failure and dysautonomia is suggested.

## 3 Cerebrospinal fluid (CSF) should be collected on presentation to exclude mimics before immunomodulatory therapy

Pleocytosis in CSF is a red flag against diagnosing GBS and may signify an infectious mimic such as Lyme disease, HIV, enterovirus or West Nile virus.[3] Elevated CSF protein with a normal white blood cell count (albuminocytological dissociation) occurs in 50%–70% of cases of GBS in the first week of disease.[3]

## 4 Intravenous immunoglobulin (IVIG) and plasma exchange (PLEX) are effective treatments that hasten recovery

The American Academy of Neurology Quality Standards Subcommittee guideline, which was supported by randomized controlled trials, recommended IVIG or PLEX for nonambulatory patients within 2 and 4 weeks of onset, respectively (Level A).[4] Both are equally effective at improving disability after 4 weeks.[4] Corticosteroids are not recommended.[4]

## 5 Nearly one-third of patients will require mechanical ventilation, but the long-term prognosis is usually favourable

Mechanical ventilation is required for almost 30% of patients with GBS and is associated with mortality of 5%–12%, slower recovery and residual disability.[5] Recovery begins after clinical nadir at 2–4 weeks, and about 83% of patients can walk independently at 6 months.[5] Persistent symptoms may include weakness in the legs, pain and fatigue.

## References

1. Sejvar JJ, Baughman AL, Wise M, et al. Population incidence of Guillain–Barré syndrome: a systematic review and meta-analysis. *Neuroepidemiology* 2011;36:123-33.
2. Baxter R, Bakshi N, Fireman B, et al. Lack of association of Guillain–Barré syndrome with vaccinations. *Clin Infect Dis* 2013;57:197-204.
3. Leonhard SE, Mandarakas MR, Gondim FAA, et al. Diagnosis and management of Guillain–Barré syndrome in ten steps. *Nat Rev Neurol* 2019;15:671-83.
4. Hughes RA, Wijdicks EF, Barohn R, et al.; Quality Standards Subcommittee of the American Academy of Neurology. Practice parameter: immunotherapy for Guillain–Barré syndrome: report of the Quality Standards Subcommittee of the American Academy of Neurology. *Neurology* 2003;23:736-40.
5. van den Berg B, Storm EF, Garssen MJP, et al. Clinical outcome of Guillain–Barré syndrome after prolonged mechanical ventilation. *J Neurol Neurosurg Psychiatry* 2018;89:949-54.

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