Clinical prognostic scales in Guillain–Barré syndrome

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The clinical presentation of Guillain–Barré syndrome (GBS) is heterogeneous and, despite effective treatments, some patients die or sustain severe disabilities. An improved clinical prognostic score for GBS facilitates early identification of patients expected to have poor outcomes. These individuals might benefit from modified treatment or participation in therapeutic trials.

Guillain–Barré syndrome (GBS) is an acute inflammatory demyelinating polyneuropathy usually triggered by an infection. Since the near-elimination of poliomyelitis, GBS is the most common cause of acute flaccid paralysis worldwide. However, the clinical presentation of GBS is heterogeneous, and existing recommendations for treatment might need to be individualized to provide the optimal therapeutic option. For this reason, clinical prognostic scales currently available for GBS have an important role in determining which treatment options are appropriate for subgroups of patients with different prognoses. As early prediction of outcome is important for both the selection of treatment options and assessing the possible benefits of these treatments, a group based at the University Medical Center in Rotterdam, The Netherlands, has improved on the clinical prognostic score they previously developed to predict the outcome of patients with GBS at an earlier point in the disease course.

...the new score can ... identify patients with GBS who have a poor prognosis at [hospital] admission...

The Erasmus GBS Outcome Score (EGOS) was the first validated prognostic indicator for GBS, which provided a simple clinical scoring system that could be applied to patients with this syndrome 2 weeks after hospital admission. EGOS relies on several variables (age, the presence of diarrhea and the disability functional score) that can accurately predict the patient’s chances of walking independently at 6 months after hospital admission. The new version developed by Walgaard et al. has improved on EGOS; the new score can now identify patients with GBS who have a poor prognosis at admission and 1 week after admission. The modified EGOS (mEGOS), as the researchers call the new score, uses similar predictive variables, such as age, the occurrence of antecedent diarrhea and the severity of GBS to assess patients at the time of admission, as well as 7 days after hospital admission. However, in mEGOS, the severity of GBS is evaluated using the Medical Research Council sum score—that is, the sum of scores (ranging from 0 [tetraplegic] to 60 [normal]) of six different muscle groups, measured bilaterally—rather than the functional grading scale used in the original EGOS. The outcomes measured were also expanded to include functional ability at 4 weeks, 3 months and 6 months after hospital admission, which is important to ensure that the new score is useful for the selection and monitoring of patients in clinical trials of new treatment regimes for patients predicted to have a poor prognosis. Consequently, mEGOS could potentially improve the clinical approach to and future management of patients with GBS who have a poor prognosis.

Current evidence from clinical trials indicates that patients who present with GBS within 2 weeks of onset of the precipitating illness and are unable to walk without help should receive either plasma exchange or intravenous immunoglobulin (IVIg), both of which will hasten the patient’s recovery and improve their outcomes. Although both treatments are effective, GBS is still associated with substantial rates of death and severe disability (between 9% and 17%). The results of some studies have also suggested that patients with GBS who have IgG antiganglioside GM1 antibodies respond better to IVIg than to plasma exchange. Patients with a poor prognosis also often have high titers of IgG antibodies against the motor gangliosides GM1, GM1b, GD1a or GalNAc-GD1a. Among patients with GBS who received IVIg as part of their treatment, those with a small rise in serum IgG levels had worse outcomes than patients who had a substantial increase in their IgG levels. In addition, serological evidence of Campylobacter jejuni or cytomegalovirus (CMV) infections are also associated with less-favorable outcomes. Patients with GBS who have a poor prognosis (that is, who have IgG antiganglioside antibodies, a small rise in serum IgG levels or serological evidence of C. jejuni or CMV) might, therefore, benefit from a second course or an increased dose of IVIg.

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Nerve conduction studies form part of the diagnostic process, and serial studies are important to define the subtype of GBS, acute inflammatory demyelinating polyneuropathy (AIDP) and acute motor axonal neuropathy (AMAN) being the two main subtypes. Evidence of inexcitable nerves and axonal degeneration on neurophysiology are associated with a poor prognosis. In a typical clinical setting, however, these laboratory tests may not be easily accessible; in such cases, mEGOS will certainly prove useful in deciding on the prognosis and treatment options for each patient. One of the potential drawbacks of mEGOS that Walgaard et al. acknowledge is its unknown utility in populations other...
than white Europeans. Studies suggest that GBS subtypes differ between Western and Asian populations. AIDP is more frequent in Western populations, whereas AMAN predominates in Asia. As mEGOS was derived from a white Dutch population, one could argue that its utility may be restricted to the AIDP subtype of GBS. However, if serial nerve conduction studies are not performed, reversible forms of AMAN (such as acute motor conduction block neuropathy) could be missed; therefore, the true incidence of AMAN in Western populations could be underestimated. Future studies of the mEGOS and EGOS in other populations will be important to clarify the validity of the model in different cohorts with GBS, and a retrospective look at the use of mEGOS and EGOS in the existing GBS databases will be important to clarify the validity of the model in these cohorts.

“Future studies … will … clarify the validity of [mEGOS] in different cohorts with GBS…”

Translating the use of these prognostic scales into practice has important implications. Both EGOS and mEGOS use clinical variables that are simple to measure. Furthermore, from a practical perspective, clinicians can use these scales at the time of hospital admission, as well as at 1 week and 2 weeks after admission, to quantify the patients’ probability of recovering the ability to walk without assistance at certain time frames. However, it should be noted that other factors, such as the neurophysiological findings and the patient’s clinical progress while on treatment, should also be taken into account. The mEGOS should greatly facilitate the identification of patients with a poor prognosis, for whom an escalation of treatment should be considered and recruitment into clinical trials assessing improved treatment modalities is vital. International collaborative efforts initiated by the International Inflammatory Neuropathy Consortium to address these issues are currently underway; these include the International GBS Outcomes Study and the International Second Dose of IVig in GBS in patients with a poor prognosis.

Competing interests
The authors declare no competing interests.