**Practice parameter: Immunotherapy for Guillain–Barré syndrome**

**Report of the Quality Standards Subcommittee of the American Academy of Neurology**

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**Abstract—Objective:** To provide an evidence-based statement to guide physicians in the management of Guillain–Barré syndrome (GBS).

**Methods:** Literature search and derivation of evidence-based statements concerning the use of immunotherapy were performed. Results: Treatment with plasma exchange (PE) or IV immunoglobulin (IVIg) hastens recovery from GBS. Combining the two treatments is not beneficial. Steroid treatment given alone is not beneficial. Recommendations: 1) PE is recommended for nonambulant adult patients with GBS who seek treatment within 4 weeks of the onset of neuropathic symptoms. PE should also be considered for ambulant patients examined within 2 weeks of the onset of neuropathic symptoms; 2) IVIg is recommended for nonambulant adult patients with GBS within 2 or possibly 4 weeks of the onset of neuropathic symptoms. The effects of PE and IVIg are equivalent; 3) Corticosteroids are not recommended for the management of GBS; 4) Sequential treatment with PE followed by IVIg, or immunoabsorption followed by IVIg is not recommended for patients with GBS; and 5) PE and IVIg are treatment options for children with severe GBS.

**Evidence review.** A search of MEDLINE from 1966 and of the Cochrane library was performed in March 2002. “Polyradiculoneuritis” was limited by “human” and cross-referenced with “therapy.” The search results were reviewed for each question by at least two members of the practice parameter group and supplemented from the reference lists in the articles retrieved and the personal reference lists of the members of the practice parameter group. Those titles representing relevant randomized controlled trials (RCTs) are included in the tables on the Neurology Web site for this article (www.neurology.org). Recommendations were graded according to the levels established by the AAN Quality Standards Subcommittee at the inception of this project (table).
Analysis of the evidence. Does initial immunotherapy hasten recovery? All studies used similar diagnostic criteria. In most, the primary outcome measure used a disability scale (0 = normal, 1 = symptoms but able to run, 2 = unable to run, 3 = unable to walk unaided, 4 = bed-bound, 5 = needing ventilation, 6 = dead). Most studies included patients with severe disease (at least grade 3 on that scale).

Plasma exchange. A Cochrane systematic review obtained data from six class II trials comparing plasma exchange (PE) alone with supportive care. The PE regimens involved exchanging approximately one plasma volume, 50 mL/kg, on five separate occasions over 1 to 2 weeks, except in one trial that used two plasma volume exchanges on alternate days for four total exchanges. One trial involving 29 participants showed a trend toward more improvement in disability after 4 weeks with PE. The other five trials showed significantly more improvement in disability grade or more patients improved in disability grade after 4 weeks. The evidence is insufficient to recommend the use of CSF filtration (level U, limited class II evidence).

Immunoabsorption. Immunoabsorption is an alternative technique to PE that removes immunoglobulins and has the advantage of not requiring the use of a human blood product as a replacement fluid. In a prospective trial with a block sequential design, there were no differences in outcome between patients treated with PE and 13 treated with immunoabsorption.

Conclusion. There is only limited class IV evidence from one small nonrandomized unblinded study.

Recommendation. The evidence is insufficient to recommend the use of immunoabsorption (level U recommendation, class IV evidence).

IV immunoglobulin. A Cochrane systematic review found no trials comparing IV Ig with placebo. In one class III trial comparing IV Ig with supportive treatment, seven of nine children who received IV Ig recovered completely by 4 weeks compared with two of nine untreated children.

Three trials compared IV Ig with PE. The mean improvement in disability grade 4 weeks after randomization was available for three trials. In a meta-analysis the weighted mean difference was 0.11 more improvement in 204 patients treated with...
In the PE-treated patients (RR, 0.11; 95% CI, 0.04 to 0.66) in the corticosteroid group (2/124, 1.6%) than in the control group (12/118, 10.2%). There was no significant difference between these regimens after 1 year. Complications were similar in the corticosteroid and placebo groups, except for hypertension, for which the RR was less (0.2; 95% CI, 0.04 to 0.66) in the corticosteroid group (2/124, 1.2%) than in the control group (12/118, 10.2%).

A comparison of a series of corticosteroid-treated patients with historical control subjects suggested a beneficial effect from corticosteroids when given in combination with IVIg. The effect of IV methylprednisolone combined with IVIg for managing GBS has been tested in a seventh randomized trial involv-
ing 233 patients, but the results have not yet been published and were not available for review.

**Conclusion.** The combined evidence from all trials shows no benefit from corticosteroids (class I evidence). The results of a trial of the combination of IV methylprednisolone and IV Ig are awaited.

**Recommendation.** Corticosteroids are not recommended for the treatment of patients with GBS (level A, class I evidence).

**Are there special issues for the treatment of children with GBS?** The clinical features of GBS in children are similar to those in adults except that severe sequelae are less common and axonal forms of the disease are more frequent in some populations. In younger children, in particular, pain is frequently the only symptom they are able to articulate, and evidence of subtle weakness and loss of reflexes may be overlooked. There is a lack of adequate randomized controlled treatment trials in children to define the role of either PE or IV Ig.

**Conclusion.** There are no adequate randomized controlled trials of treatment in children.

**Recommendation.** PE and IV Ig are treatment options for children with severe GBS (level B recommendation derived from class II evidence in adults).

**Future research.** More research is needed to evaluate immunotherapy for patients with GBS, particularly the use of combination treatments and further treatment after the initial course, especially for those patients who do not respond. There is a need to identify patients who are at greater risk of an adverse outcome and to discover whether subgroups, including children, and people with axonal forms of GBS and Fisher’s syndrome have differential responses to treatment. Research should also investigate the best methods of supportive care for monitoring autonomic and pulmonary function, weaning from ventilation, treating pain, managing fatigue, and rehabilitation.

**Disclaimer.** This statement is provided as an educational service of the American Academy of Neurology. It is based on an assessment of current scientific and clinical information. It is not intended to include all possible proper methods of care for a particular neurologic problem or all legitimate criteria for choosing to use a specific procedure. Neither is it intended to exclude any reasonable alternative methodologies. The AAN recognizes that specific patient care decisions are the prerogative of the patient and the physician caring for the patient, based on all of the circumstances involved.

**Acknowledgment**

The GBS Practice Parameter Group thank Drs. D. Annane, S. Chevret, F.G.A. van der Meché, P.A. van Doorn, J.C. Raphael, and A.V. Swan, the authors of the Cochrane systematic reviews on which this practice parameter is in part based, Dr. Allan Ropper, and the Guillain–Barré Syndrome Foundation International for assistance.

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Neurology 2003;61:736-740
DOI 10.1212/WNL.61.6.736

This information is current as of September 22, 2003