EVIDENCE-BASED GUIDELINE: IV IMMUNOGLOBULIN IN THE TREATMENT OF NEUROMUSCULAR DISORDERS: REPORT OF THE THERAPEUTICS AND TECHNOLOGY ASSESSMENT SUBCOMMITTEE OF THE AMERICAN ACADEMY OF NEUROLOGY

Hai-feng Li, Xiang Gao, Yu Hong, Qingdao, China: The American Academy of Neurology (AAN) guideline on IV immunoglobulin (IVIg) for neuromuscular diseases recommended that IVIg should be considered in the treatment of myasthenia gravis (MG).[1] The evidence is based from studies on patients with MG with worsening weakness.

We agree that IVIg is probably effective in treating both acute exacerbations of MG and severe MG. This is consistent with 2 European Federation of Neurological Societies guidelines,[2,3] an American Association of Neuromuscular & Electrodiagnostic Medicine consensus statement,[4] and a Cochrane review.[5] We are facing other challenges with regard to MG. IVIg is often used to prepare patients with MG for thymectomy or other types of surgery. It is also used for severe MG or MG exacerbations during pregnancy. There are no controlled studies for these practices. However, the short-term effect of IVIg in acute exacerbations and its safety profile suggests IVIg is useful in these situations.[2] Existing data are insufficient to support the efficacy of repeated use of IVIg in MG in improving functional outcome or steroid-sparing effect.[2–5]

Considering the short-term effects of a single course of IVIg and the cost-effectiveness of long-term IVIg treatment, we believe the recommendation of this AAN guideline is oversimplified and more detailed explanations are necessary.

Author Response: Huned S. Patwa, New Haven, CT; Vinay Chaudhry, Baltimore; Alex D. Rae-Grant, Cleveland; Yuen T. So, Palo Alto, CA: We thank Drs. Li et al. for their comments regarding our article.[1] As the authors indicate, our findings are consistent with those from other guidelines addressing the same topic, although different methodologies may result in different recommendations. Our analysis is based on the process outlined in the AAN guideline development manual.[6] The evidence supports a Level B recommendation for moderately severe MG and the details can be found in the evidence tables. There are no adequate studies to address the efficacy of a single course of IVIg for the treatment of acute exacerbations of MG. As indicated in our clinical context section, further studies are warranted, as there are few adequately powered, randomized studies for the use of IVIg in MG.

1. Patwa HS, Chaudhry V, Katzberg H, Rae-Grant AD, So YT. Evidence-based guideline: intravenous immunoglobulin in the treatment of neuromuscular disorders: report of the Therapeutics and Technology Assessment Subcommittee of the American Academy of Neurology. Neurology 2012; 78:1009–1015.
2. Elovaara I, Apostolski S, van Doorn P, et al. EFNS guidelines for the use of intravenous immunoglobulin in treatment of neurological diseases: EFNS task force on the use of intravenous immunoglobulin in treatment of neurological diseases. Eur J Neurol 2008;15:893–908.
3. Skeie GO, Apostolski S, Evoli A, et al. Guidelines for treatment of autoimmune neuromuscular transmission disorders. Eur J Neurol 2010;17:893–902.
4. Donofrio PD, Berger A, Brannagan TH 3rd, et al. Consensus statement: the use of intravenous immunoglobulin in the treatment of neuromuscular conditions report of the AANEM ad hoc committee. Muscle Nerve 2009;40:890–900.
5. Gajdos P, Chevret S, Toyka K. Intravenous immunoglobulin for myasthenia gravis. Cochrane Database Syst Rev 2008; CD002277.
6. Edlund W, Gronseth G, So Y, Franklin G. AAN Clinical Practice Guideline Process Manual. St. Paul: American Academy of Neurology; 2005.

CORRECTION

Patient-reported impact of symptoms in myotonic dystrophy type 1 (PRISM-1)

In the article "Patient-reported impact of symptoms in myotonic dystrophy type 1 (PRISM-1)" by C. Heatwole et al. (*Neurology*® 2012;79:348–357), Nevada was omitted from footnote b in table 1. The authors regret the omission.

Author disclosures are available upon request (journal@neurology.org).