

Disputes & Debates: Editors' Choice

Steven Galetta, MD, FAAN, Section Editor

Editors' note: Practice guideline update recommendations summary: Disorders of consciousness: Report of the Guideline Development, Dissemination, and Implementation Subcommittee of the American Academy of Neurology; the American Congress of Rehabilitation Medicine; and the National Institute on Disability, Independent Living, and Rehabilitation Research

In their American Academy of Neurology (AAN) practice parameter, Giacino et al. provided a thorough review of the available evidence pertaining to the care of patients with impaired consciousness. The expert panel provided level of recommendations (LORs) regarding the discussion of long-term care needs, pain management strategies, and techniques for neuroprognostication in patients with disorders of consciousness. In response to these consensus recommendations, Phan et al. highlight 1 potential limitation of the LOR classification system that was used. Historically, the highest LOR (level A) was afforded only to recommendations based on 1 or more randomized clinical trials. However, this requirement was amended by the Institute of Medicine in 2011 as well as the 2011 AAN Clinical Guideline Practice Manual, as the authors emphasize in their response. After 2011, a level A recommendation was permitted as long as there was strong and consistent related evidence and inferences could be drawn. Therefore, a higher LOR could be assigned to recommendations with less explicit substantiation from large randomized clinical trials. By using this classification schema, some recommendations may be generalized to patients who are likely to benefit from such guidance.

James E. Siegler III, MD, and Steven Galetta, MD
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Reader response: Practice guideline update recommendations summary: Disorders of consciousness: Report of the Guideline Development, Dissemination, and Implementation Subcommittee of the American Academy of Neurology; the American Congress of Rehabilitation Medicine; and the National Institute on Disability, Independent Living, and Rehabilitation Research

Thanh G. Phan (Clayton, Australia), Udaya Seneviratne (Clayton, Australia), and Henry Ma (Clayton, Australia)
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We read with interest the disorders of consciousness guideline¹ but found issues with the recommendations. Some of the recommendations are classified as level A (recommendations 3, 9, and 11). For example, “When prognosis is poor, long-term care must be discussed (level A)...”¹ The references cited did not come from a randomized control trial. Typically, level A is based on one or more randomized control trial and is prefaced by a statement about the class of evidence. We cannot find references to any trials on which these recommendations were made.^{1,2} Can the authors reassess the use of the level of recommendation in this guideline?

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

1. Giacino JT, Katz DI, Schiff ND, et al. Practice guideline update recommendations summary: disorders of consciousness: report of the Guideline Development, Dissemination, and Implementation Subcommittee of the American Academy of Neurology; the American Congress of Rehabilitation Medicine; and the National Institute on Disability, Independent Living, and Rehabilitation Research. *Neurology* 2018;91:450–460.
2. Giacino JT, Katz DI, Schiff ND, et al. Comprehensive systematic review update summary: disorders of consciousness: report of the Guideline Development, Dissemination, and Implementation Subcommittee of the American Academy of Neurology; the American Congress of Rehabilitation Medicine; and the National Institute on Disability, Independent Living, and Rehabilitation Research. *Neurology* 2018;91:461–470.

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Author response: Practice guideline update recommendations summary: Disorders of consciousness: Report of the Guideline Development, Dissemination, and Implementation Subcommittee of the American Academy of Neurology; the American Congress of Rehabilitation Medicine; and the National Institute on Disability, Independent Living, and Rehabilitation Research

Melissa J. Armstrong (Gainesville, FL), Joseph T. Giacino (Boston), Douglas I. Katz (Braintree, MA), Nicholas D. Schiff (New York), John Whyte (Elkins Park, PA), Eric J. Ashman (Kalamazoo, MI), Stephen Ashwal (Loma Linda, CA), Richard Barbano (Rochester, NY), Flora M. Hammond (Indianapolis), Steven Laureys (Liège, Belgium), Geoffrey S.F. Ling (Baltimore), Risa Nakase-Richardson (Tampa, FL), Ronald T. Seel (Richmond, VA), Stuart Yablon (Jackson, MS), Thomas S.D. Getchius (Washington, DC), and Gary S. Gronseth (Kansas City, KS) *Neurology*® 2019;92:1164. doi:10.1212/WNL.0000000000007669

American Academy of Neurology (AAN) guidelines comply with the AAN Institute Board-approved guideline methodology referenced within the systematic review/guideline.^{1,2} Compliance is ensured by a methodologist working on each project and multiple rounds of AAN Guideline Development, Dissemination, and Implementation Subcommittee review. We believe that Phan et al. are referencing the 2004 recommendation methodology.³ The disorders of consciousness guideline used the 2011 AAN guideline manual, as amended,⁴ based on 2011 Institute of Medicine (IOM) standards for evidence-based guidelines.⁵ In this process, recommendations are based not only on a systematic review of the evidence but also on strongly related evidence, principles of care, and inferences. The level of obligation for each recommendation is determined by the strength of these premises and a risk–benefit assessment, with adjustments based on outcome importance, patient preference variability, feasibility/availability, and patient costs. Consensus is determined by a modified Delphi voting process in accordance with prespecified rules, as described in the systematic review.² This IOM-compliant approach improves recommendation usability. The modified Delphi tables and the premise types for each recommendation rationale are available in the online appendices, NPub.org/m5ii8i (“rationale profiles” for recommendations 3, 9, and 11 are on pages 190, 204, and 206, respectively).

1. Giacino JT, Katz DI, Schiff ND, et al. Practice guideline update recommendations summary: disorders of consciousness: report of the Guideline Development, Dissemination, and Implementation Subcommittee of the American Academy of Neurology; the American Congress of Rehabilitation Medicine; and the National Institute on Disability, Independent Living, and Rehabilitation Research. *Neurology* 2018;91:450–460.
2. Giacino JT, Katz DI, Schiff ND, et al. Comprehensive systematic review update summary: disorders of consciousness: report of the Guideline Development, Dissemination, and Implementation Subcommittee of the American Academy of Neurology; the American Congress of Rehabilitation Medicine; and the National Institute on Disability, Independent Living, and Rehabilitation Research. *Neurology* 2018;91:461–470.
3. American Academy of Neurology. Clinical Practice Guideline Process Manual, 2004 ed. St. Paul, MN: American Academy of Neurology; 2004.
4. American Academy of Neurology. Clinical Practice Guideline Process Manual, 2011 ed. St. Paul: American Academy of Neurology; 2011.
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Author disclosures are available upon request (journal@neurology.org).

Editors' note: Clinical Reasoning: A 54-year-old woman with confusion and visual disturbances

Rossi et al. presented the unusual case of a 54-year-old woman with cirrhosis who developed oculomotor apraxia, optic ataxia, impaired smooth pursuit, and horizontal nystagmus in all directions of gaze. The neuroimaging and electrographic diagnosis was nonconvulsive status epilepticus resulting in Bálint syndrome. Dr. Pollak also suspects an epileptic origin of the horizontal, alternating nystagmus pattern, given the bilateral MRI and EEG findings. However, Dr. Pollack notes that a normal optokinetic nystagmus would be unusual during seizure activity. Rossi et al. attribute this to the fluctuating nature of the patient's condition and the intermittent epileptiform activity on EEG. Resolution of the cortical diffusion abnormalities on MRI would also have supported seizures as the cause of the patient's symptoms, as Dr. Pollak writes. Unfortunately, this could not be confirmed as the patient was lost to follow-up.

James E. Siegler III, MD, and Steven Galetta, MD
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Reader response: Clinical Reasoning: A 54-year-old woman with confusion and visual disturbances

Lea Pollak (Ness Ziona, Israel)
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In the Resident & Fellow Clinical Reasoning paper by Rossi et al.,¹ the authors described an unusual case of Bálint syndrome caused by focal nonconvulsive status epilepticus in a patient with cirrhosis and hyponatremia. I am curious about the nature of the clinical finding: "... horizontal nystagmus in all directions including on primary gaze."¹

Horizontal nystagmus in all directions localizes to the brainstem/cerebellum; however, in this case,¹ the lesions were parieto-occipital. Hyponatremia, if accompanied by hypomagnesemia, would cause a downbeat nystagmus. Could the nystagmus thus be an epileptic nystagmus of cortical origin? The bilaterality of the epileptic foci might explain the bilateral direction of the nystagmus. The authors describe an intermittent eye deviation on video during EEG recording; the mechanism is, therefore, probably due to epileptic alternative eye deviation with quick corrective saccades. It would be interesting to know the direction of the nystagmus, since this may elucidate whether the underlying activated mechanism of the eye deviations was saccadic or pursuit. Furthermore, the finding of a normal optokinetic nystagmus in Bálint syndrome and during seizures is mostly unusual. Also, can the authors please comment on the radiologic follow-up of this patient as the parieto-occipital T2 hyperintensities should resolve with time if attributed to seizure activity?

1. Rossi KC, Brandstadter R, Fields MC, Leong J, Shin S. Clinical Reasoning: a 54-year-old woman with confusion and visual disturbances. *Neurology* 2018;91:363–367.

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Author response: Clinical Reasoning: A 54-year-old woman with confusion and visual disturbances

Kyle C. Rossi (New York), Rachel Brandstadter (New York), Madeline C. Fields (New York), and Susan Shin (New York)
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We thank Dr. Pollak for the thoughtful comments on our article.¹ The nature of the nystagmus was variable over the clinical course. Our earliest notes described direction-changing horizontal gaze-evoked nystagmus on left and right end gaze and primary gaze. The mechanism of epileptic nystagmus is poorly understood with most available literature being from case reports, often reporting the fast phase of nystagmus away from the seizure focus.^{2–4} Here, the bilateral foci could explain the direction changing nature of the nystagmus. Of note, the case was confounded by metabolic derangements, potentially contributing to brainstem dysfunction and eye movement abnormalities. Although epileptic nystagmus is possible, it is difficult to conclude with certainty.

The intact optokinetic nystagmus (OKN) reflex could be related to the fluctuating nature of the symptoms given an epileptic origin as opposed to a fixed structural origin. Additionally, Baloh et al.⁵ reported on the structural pathways involved in the OKN reflex, suggesting a complicated 2-pathway mechanism and showing that many parietal lesions do not obliterate all parts of the OKN response uniformly.

Regarding follow-up imaging, the patient was unfortunately lost to follow-up from a neurology perspective; the plan for follow-up imaging was not completed at our institution.

1. Rossi KC, Brandstadter R, Fields MC, Leong J, Shin S. Clinical Reasoning: a 54-year-old woman with confusion and visual disturbances. *Neurology* 2018;91:363–367.
2. Lee SU, Suh HI, Choi JY, et al. Epileptic nystagmus: a case report and systematic review. *Epilepsy Behav Case Rep* 2014;2:156–160.
3. Ma Y, Wang J, Li D, Lang S. Two types of isolated epileptic nystagmus: case report. *Int J Clin Exp Med* 2015;8:13500–13507.
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CORRECTION

Quality of life predicts outcome of deep brain stimulation in early Parkinson disease

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In the article “Quality of life predicts outcome of deep brain stimulation in early Parkinson disease” by Schuepbach et al.,¹ published online ahead of print on February 8, 2019, Dr. Hälbig’s name should have included a middle initial: Thomas D. Hälbig. The corrected name appears in the March 5 issue. The editorial office regrets the error.

Reference

1. Schuepbach WMM, Tonder L, Schnitzler A, et al. Quality of life predicts outcome of deep brain stimulation in early Parkinson disease. *Neurology* 2019;92:e1109–e1120.

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Quality of life predicts outcome of deep brain stimulation in early Parkinson disease

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