General

Guideline Title


Bibliographic Source(s)


Guideline Status

This is the current release of the guideline.


The American Academy of Neurology reaffirmed the currency of this guideline in 2013.

Regulatory Alert

FDA Warning/Regulatory Alert

Note from the National Guideline Clearinghouse: This guideline references a drug(s) for which important revised regulatory and/or warning information has been released.

- August 31, 2016 – Opioid pain and cough medicines combined with benzodiazepines: A U.S. Food and Drug Administration (FDA) review has found that the growing combined use of opioid medicines with benzodiazepines or other drugs that depress the central nervous system (CNS) has resulted in serious side effects, including slowed or difficult breathing and deaths. FDA is adding Boxed Warnings to the drug labeling of prescription opioid pain and prescription opioid cough medicines and benzodiazepines.
- March 22, 2016 – Opioid pain medicines: The U.S. Food and Drug Administration (FDA) is warning about several safety issues with the entire class of opioid pain medicines. These safety risks are potentially harmful interactions with numerous other medications, problems with the adrenal glands, and decreased sex hormone levels. They are requiring changes to the labels of all opioid drugs to warn about these risks.
Recommendations

Major Recommendations

Definitions of the levels of the recommendations (A, B, C, U) and classification of the evidence (Class I-IV) are provided at the end of the "Major Recommendations" field.

Breaking the News

How should a physician tell patients that they have amyotrophic lateral sclerosis (ALS)?

Conclusion

There have been no controlled trials of breaking the news in ALS.

Recommendation

There is insufficient evidence to support or refute any specific method of disclosing the diagnosis in ALS (Level U).

Multidisciplinary Clinic

Does multidisciplinary management improve outcomes?

Conclusions

Two Class II studies and 1 Class III study show that multidisciplinary clinics specializing in ALS care are probably effective in several ways: increased use of adaptive equipment; increased utilization of riluzole, percutaneous endoscopic gastrostomy (PEG), and noninvasive ventilation (NIV); improved quality of life; and lengthened survival. However, 1 Class II study with low use of treatments found no survival benefit.

Recommendations

Specialized multidisciplinary clinic referral should be considered for patients with ALS to optimize health care delivery (Level B) and prolong survival (Level B), and may be considered to enhance quality of life (Level C).

Symptomatic Management

What are the most effective treatments for sialorrhea?

Conclusions

In patients with medically refractory sialorrhea, botulinum toxin B (BTxB) injections into the parotid and submandibular glands are probably effective (1 Class I study). There are inadequate data on the effectiveness of botulinum toxin A (BTxA) and amitriptyline (1 Class III study). Low-dose irradiation is possibly effective for sialorrhea (2 Class III studies).

Recommendations

In patients with ALS who have medically refractory sialorrhea, BTxB should be considered (Level B) and low-dose radiation therapy to the salivary glands may be considered (Level C).

What pharmacologic measures reduce pseudobulbar affect?

Conclusions

The combination of dextromethorphan/quinidine (DM/Q) is probably effective for pseudobulbar affect in ALS (1 Class I study), although side effects may limit its usefulness.

Recommendation

If approved by the Food and Drug Administration (FDA), and if side effects are acceptable, DM/Q should be considered for symptoms of pseudobulbar affect in patients with ALS (Level B).

What pharmacologic interventions reduce fatigue?
Conclusions

There are no controlled studies of pharmacologic agents relieving fatigue in ALS. Riluzole possibly causes fatigue in some patients (2 Class III studies).

Recommendations

In patients developing fatigue while taking riluzole, once risks of fatigue versus modest survival benefits have been discussed, withholding the drug may be considered (Level C).

What interventions reduce cramps?

Conclusions

Studies of gabapentin, vitamin E, and riluzole for treating cramps were all negative (Class III). There are safety concerns about quinine.

Recommendations

There are insufficient data to support or refute any specific intervention for the treatment of cramps in ALS (Level U).

What interventions reduce spasticity?

Conclusion

Evidence is insufficient to recommend exercise or medication for treating spasticity in ALS (Class III).

Recommendation

There are insufficient data to support or refute exercise or medication for treating spasticity in ALS (Level U).

What pharmacologic interventions reduce depression?

Conclusion

There have been no controlled trials of treatment for depression in ALS.

Recommendation

There are insufficient data to support or refute specific treatments for depression in ALS (Level U).

What pharmacologic interventions reduce anxiety?

Conclusion

There have been no trials of treatment for anxiety in ALS.

Recommendation

There are insufficient data to support or refute specific treatment for anxiety in ALS (Level U).

What pharmacologic interventions reduce insomnia?

Conclusion

There have been no studies of treatment for insomnia in ALS.

Recommendation

There are insufficient data to support or refute specific treatment for insomnia in ALS (Level U).

Cognitive and Behavioral Impairment

What is the prevalence and natural history of cognitive and behavioral impairment in ALS?

Conclusions
A significant proportion of patients with ALS demonstrate cognitive impairment and some have dementia (2 Class II, multiple Class III studies). Neither behavioral impairment in ALS nor the natural progression of cognitive or behavioral impairments has been adequately studied.

**Recommendation**

Screening for cognitive and behavioral impairment should be considered in patients with ALS (Level B).

How is cognitive or behavioral impairment in ALS diagnosed?

**Conclusion**

Neuropsychological assessment is possibly effective for identifying cognitive impairment in ALS (1 Class II, 1 Class III).

**Recommendation**

Screening tests of executive function may be considered to detect cognitive impairment in patients with ALS prior to confirmation with formal neuropsychological evaluation (Level C).

What is the effect of cognitive or behavioral impairment on management of patients with ALS?

**Conclusion**

Insufficient data exist on the effect of cognitive or behavioral impairment on the management of patients with ALS.

**Recommendation**

There are insufficient data to support or refute the impact of cognitive and behavioral impairment on management in ALS (Level U).

What treatments are effective for cognitive or behavioral impairment in ALS?

**Conclusions**

Data are inadequate regarding the effect of pharmacologic treatment or NIV for cognitive or behavioral impairment in ALS.

**Recommendation**

There are insufficient data to support or refute treatment of cognitive or behavioral impairment in ALS (Level U).

**Communication**

What treatments for dysarthria optimize communication in ALS?

**Conclusion**

No controlled studies examined communication in ALS.

**Recommendation**

There are insufficient data to support or refute treatment to optimize communication in ALS (Level U).

**Palliative Care**

What treatments reduce pain and dyspnea in the terminal phase of ALS?

**Conclusion**

No controlled studies examined treating pain or dyspnea in late-stage ALS.

**Recommendation**

There are insufficient data to support or refute specific treatments for pain and dyspnea in late-stage ALS (Level U).

Do hospice care, spiritual interventions, or advance directives improve quality of life in the terminal phase of ALS?

**Conclusion**
No controlled studies examined hospice, spiritual care, or advance directives in ALS.

**Recommendation**

There are insufficient data to support or refute hospice, spiritual care, or advance directives in ALS (Level U).

What is the optimal method of withdrawing both noninvasive and invasive ventilation in ALS?

**Conclusion**

There are no controlled studies examining withdrawal of ventilation in ALS.

**Recommendation**

There are insufficient data to support or refute specific strategies for withdrawal of ventilation in ALS (Level U).

**Definitions:**

**Classification of Evidence for Studies of Therapeutic Intervention**

Class I = A randomized, controlled clinical trial of the intervention of interest with masked or objective outcome assessment, in a representative population. Relevant baseline characteristics are presented and substantially equivalent among treatment groups or there is appropriate statistical adjustment for differences. The following are also required:

a. Concealed allocation
b. Primary outcome(s) clearly defined
c. Exclusion/inclusion criteria clearly defined
d. Adequate accounting for drop-outs (with at least 80% of enrolled subjects completing the study) and cross-overs with numbers sufficiently low to have minimal potential for bias
e. For non-inferiority or equivalence trials claiming to prove efficacy for one or both drugs, the following are also required*:
   1. The authors explicitly state the clinically meaningful difference to be excluded by defining the threshold for equivalence or non-inferiority.
   2. The standard treatment used in the study is substantially similar to that used in previous studies establishing efficacy of the standard treatment. (e.g., for a drug, the mode of administration, dose and dosage adjustments are similar to those previously shown to be effective).
   3. The inclusion and exclusion criteria for patient selection and the outcomes of patients on the standard treatment are comparable to those of previous studies establishing efficacy of the standard treatment.
   4. The interpretation of the results of the study is based upon a per protocol analysis that takes into account dropouts or crossovers.

Class II = A randomized controlled clinical trial of the intervention of interest with masked or objective outcome assessment that lacks one criteria a–e above or a prospective matched cohort study with masked or objective outcome assessment in a representative population that meets b–e above. Relevant baseline characteristics are presented and substantially equivalent among treatment groups or there is appropriate statistical adjustment for differences.

Class III = All other controlled trials (including well-defined natural history controls or patients serving as own controls) in a representative population, where outcome is independently assessed, or independently derived by objective outcome measurement.**

Class IV = Studies not meeting Class I, II, or III criteria including consensus or expert opinion.

*Note that numbers 1-3 in Class Ie are required for Class II in equivalence trials. If any one of the three is missing, the class is automatically downgraded to Class III.

**Objective outcome measurement: an outcome measure that is unlikely to be affected by an observer's (patient, treating physician, investigator) expectation or bias (e.g., blood tests, administrative outcome data).

**Classification of Evidence for Diagnostic Accuracy**

Class I = A cohort study with prospective data collection of a broad spectrum of persons with the suspected condition, using an acceptable reference standard for case definition. The diagnostic test is objective or performed and interpreted without knowledge of the patient's clinical status. Study results allow calculation of measures of diagnostic accuracy.

Class II = A case control study of a broad spectrum of persons with the condition established by an acceptable reference standard compared to a broad spectrum of controls or a cohort study where a broad spectrum of persons with the suspected condition where the data was collected
The diagnostic test is objective or performed and interpreted without knowledge of the disease status. Study results allow calculation of measures of diagnostic accuracy.

Class III = A case control study or a cohort study where either the persons with the condition or the controls are of a narrow spectrum. The condition is established by an acceptable reference standard. The reference standard and diagnostic test are objective or performed and interpreted by different observers. Study results allow calculation of measures of a diagnostic accuracy.

Class IV = Studies not meeting Class I, II, or III criteria including consensus, expert opinion or a case report.

Classification of Recommendations

Level A = Established as effective, ineffective, or harmful (or established as useful/predictive or not useful/predictive) for the given condition in the specified population. (Level A rating requires at least two consistent Class I studies.)*

Level B = Probably effective, ineffective, or harmful (or probably useful/predictive or not useful/predictive) for the given condition in the specified population. (Level B rating requires at least one Class I study or two consistent Class II studies.)

Level C = Possibly effective, ineffective, or harmful (or possibly useful/predictive or not useful/predictive) for the given condition in the specified population. (Level C rating requires at least one Class II study or two consistent Class III studies.)

Level U = Data inadequate or conflicting; given current knowledge, treatment (test, predictor) is unproven.

*In exceptional cases, one convincing Class I study may suffice for an "A" recommendation if: 1) all criteria are met, 2) the magnitude of effect is large (relative rate improved outcome >5 and the lower limit of the confidence interval is >2).

Clinical Algorithm(s)

None provided

Scope

Disease/Condition(s)

Amyotrophic lateral sclerosis (ALS; also known as Lou Gehrig's disease)

Guideline Category

Assessment of Therapeutic Effectiveness

Evaluation

Management

Risk Assessment

Screening

Treatment

Clinical Specialty

Family Practice

Internal Medicine

Neurology
Psychiatry

Psychology

Intended Users

Advanced Practice Nurses
Nurses
Occupational Therapists
Physical Therapists
Physician Assistants
Physicians
Psychologists/Non-physician Behavioral Health Clinicians
Respiratory Care Practitioners
Social Workers
Speech-Language Pathologists

Guideline Objective(s)

To systematically review evidence bearing on the management of patients with amyotrophic lateral sclerosis

Target Population

Patients with amyotrophic lateral sclerosis

Interventions and Practices Considered

1. Communicating the diagnosis to the patient
2. Multidisciplinary team approach
   - Physician
   - Physical therapist
   - Occupational therapist
   - Speech pathologist
   - Dietitian
   - Social worker
   - Respiratory therapist
   - Nurse case manager
3. Management of refractory sialorrhea
   - Botulinum toxin type B injections into the parotid and submandibular glands
   - Low-dose radiation therapy
   - Management of pseudobulbar effects (emotional lability)
   - A fixed-dose combination of dextromethorphan/quinidine
   - Screening for cognitive and behavioral impairment

Major Outcomes Considered
Methodology

Methods Used to Collect/Select the Evidence

Searches of Electronic Databases

Description of Methods Used to Collect/Select the Evidence

2009 Guideline

The authors searched OVID, MEDLINE, EMBASE, CINAHL, Science Citation Index, BIOETHICSLINE, International Pharmaceutical Abstracts, OVID Current Contents, Medline-Proquest, EIIFL, and INVEST from 1998 through September 2007 combining the words ALS, Lou Gehrig's disease, and motor neuron disease with the following words using AND: sialorrhea, pseudobulbar palsy, pseudobulbar affect, emotional lability, palliative care, diagnosis, telling the diagnosis, breaking the news, advance directives, botulinum toxin A, botulinum toxin B, parotid irradiation, anticholinergic drugs, amitriptyline, glycopyrrolate, benztropine, transdermal hyoscymine, atropine, trihexyphenidyl hydrochloride, propranolol, metoprolol, dextromethorphan, quinidine, opioids, opiates, oxygen, hospice, dyspnea, pain, lorazepam, anxiety, sleep, depression, cramps, spasticity, insomnia, deep venous thrombosis, communication devices, multidisciplinary clinic, specialty clinic, cognitive impairment, dementia, frontotemporal dementia, executive dysfunction, fatigue, and constipation. The authors reviewed the abstracts of these articles and examined 142 articles in their entirety.

2013 Reaffirmation

The guideline developer searched OVID MEDLINE, EMBASE, CINAHL, Science Citation Index, BIOETHICSLINE, International Pharmaceutical Abstracts (IPAB), OVID Current contents, Medline-ProQuest, EIIFL, and INVEST for studies published between 2010 and 2013 using the following search terms: ALS, Lou Gehrig's disease, and motor neuron disease with the following words using AND: sialorrhea, pseudobulbar palsy, pseudobulbar affect, emotional lability, palliative care, diagnosis, telling the diagnosis, breaking the news, advance directives, botulinum toxin A, botulinum toxin B, parotid irradiation, anticholinergic drugs, amitriptyline, glycopyrrolate, benztropine, transdermal hyoscymine, atropine, trihexyphenidyl hydrochloride, propranolol, metoprolol, dextromethorphan, quinidine, opioids, opiates, oxygen, hospice, dyspnea, pain, lorazepam, anxiety, sleep, depression, cramps, spasticity, insomnia, deep venous thrombosis, communication devices, multidisciplinary clinic, specialty clinic, cognitive impairment, dementia, frontotemporal dementia, executive dysfunction, fatigue, and constipation.

Number of Source Documents

Not stated

Methods Used to Assess the Quality and Strength of the Evidence

Expert Consensus

Weighting According to a Rating Scheme (Scheme Given)

Rating Scheme for the Strength of the Evidence

Classification of Evidence for Studies of Therapeutic Intervention

Class I = A randomized, controlled clinical trial of the intervention of interest with masked or objective outcome assessment, in a representative population. Relevant baseline characteristics are presented and substantially equivalent among treatment groups or there is appropriate statistical
adjustment for differences. The following are also required:

a. Concealed allocation
b. Primary outcome(s) clearly defined
c. Exclusion/inclusion criteria clearly defined
d. Adequate accounting for drop-outs (with at least 80% of enrolled subjects completing the study) and cross-overs with numbers sufficiently low to have minimal potential for bias
e. For non-inferiority or equivalence trials claiming to prove efficacy for one or both drugs, the following are also required*:
   1. The authors explicitly state the clinically meaningful difference to be excluded by defining the threshold for equivalence or non-inferiority.
   2. The standard treatment used in the study is substantially similar to that used in previous studies establishing efficacy of the standard treatment. (e.g., for a drug, the mode of administration, dose and dosage adjustments are similar to those previously shown to be effective).
   3. The inclusion and exclusion criteria for patient selection and the outcomes of patients on the standard treatment are comparable to those of previous studies establishing efficacy of the standard treatment.
   4. The interpretation of the results of the study is based upon a per protocol analysis that takes into account dropouts or crossovers.

Class II = A randomized controlled clinical trial of the intervention of interest in a representative population with masked or objective outcome assessment that lacks one criteria a–e above or a prospective matched cohort study with masked or objective outcome assessment in a representative population that meets b–e above. Relevant baseline characteristics are presented and substantially equivalent among treatment groups or there is appropriate statistical adjustment for differences.

Class III = All other controlled trials (including well-defined natural history controls or patients serving as own controls) in a representative population, where outcome is independently assessed, or independently derived by objective outcome measurement.**

Class IV = Studies not meeting Class I, II, or III criteria including consensus or expert opinion.

*Note that numbers 1-3 in Class Ie are required for Class II in equivalence trials. If any one of the three is missing, the class is automatically downgraded to Class III.

**Objective outcome measurement: an outcome measure that is unlikely to be affected by an observer's (patient, treating physician, investigator) expectation or bias (e.g., blood tests, administrative outcome data).

Classification of Evidence for Diagnostic Accuracy

Class I = A cohort study with prospective data collection of a broad spectrum of persons with the suspected condition, using an acceptable reference standard for case definition. The diagnostic test is objective or performed and interpreted without knowledge of the patient's clinical status. Study results allow calculation of measures of diagnostic accuracy.

Class II = A case control study of a broad spectrum of persons with the condition established by an acceptable reference standard compared to a broad spectrum of controls or a cohort study where a broad spectrum of persons with the suspected condition where the data was collected retrospectively. The diagnostic test is objective or performed and interpreted without knowledge of the disease status. Study results allow calculation of measures of diagnostic accuracy.

Class III = A case control study or a cohort study where either the persons with the condition or the controls are of a narrow spectrum. The condition is established by an acceptable reference standard. The reference standard and diagnostic test are objective or performed and interpreted by different observers. Study results allow calculation of measures of a diagnostic accuracy.

Class IV = Studies not meeting Class I, II, or III criteria including consensus, expert opinion or a case report.

Methods Used to Analyze the Evidence

Systematic Review with Evidence Tables

Description of the Methods Used to Analyze the Evidence

The diagnostic and therapeutic classification schemes used to grade the articles are summarized in "Rating Scheme for the Strength of the Evidence" field.
Methods Used to Formulate the Recommendations

Expert Consensus

Description of Methods Used to Formulate the Recommendations

2009 Guideline

The American Academy of Neurology (AAN) assembled a panel of experts with expertise in amyotrophic lateral sclerosis (ALS). The Quality Standards Subcommittee of the American Academy of Neurology developed a set of clinical questions relevant to the evaluation of multidisciplinary care, symptom management, and cognitive/behavioral impairment issues related to the care of patients with ALS. The strength of the practice recommendations was directly linked to the class of evidence using the scheme described in the "Rating Scheme for the Strength of the Recommendations" field.

2013 Reaffirmation

The AAN assesses their clinical practice guidelines every 2 years to determine whether new literature has been published that would warrant an update. The following steps are taken:

- Biennial correspondence is sent to all authors and the facilitator.
- An updated literature search and a review of methodological soundness are performed by a Guideline Development Subcommittee (GDS) member. (Note: The search should specifically seek to identify new evidence that would change the conclusions in the systematic review or recommendations in the CPG.)

All documents biennially reviewed by the GDS that don't require an update are reaffirmed. See the AAN Clinical Practice Guideline Process Manual for additional information.

Rating Scheme for the Strength of the Recommendations

Classification of Recommendations

Level A = Established as effective, ineffective, or harmful (or established as useful/predictive or not useful/predictive) for the given condition in the specified population. (Level A rating requires at least two consistent Class I studies.*)

Level B = Probably effective, ineffective, or harmful (or probably useful/predictive or not useful/predictive) for the given condition in the specified population. (Level B rating requires at least one Class I study or two consistent Class II studies.)

Level C = Possibly effective, ineffective, or harmful (or possibly useful/predictive or not useful/predictive) for the given condition in the specified population. (Level C rating requires at least one Class II study or two consistent Class III studies.)

Level U = Data inadequate or conflicting; given current knowledge, treatment (test, predictor) is unproven.

*In exceptional cases, one convincing Class I study may suffice for an "A" recommendation if: 1) all criteria are met, 2) the magnitude of effect is large (relative rate improved outcome >5 and the lower limit of the confidence interval is >2).

Cost Analysis

A formal cost analysis was not performed and published cost analyses were not reviewed.

Method of Guideline Validation

External Peer Review

Internal Peer Review

Description of Method of Guideline Validation
The guidelines were approved by the American Academy of Neurology (AAN) Quality Standards Subcommittee on November 5, 2008, by the Practice Committee on March 8, 2009, and by the Executive Board of the American Academy of Neurology on July 30, 2009.

Evidence Supporting the Recommendations

Type of Evidence Supporting the Recommendations

The type of supporting evidence is identified and graded for each recommendation (see the "Major Recommendations" field).

Benefits/Harms of Implementing the Guideline Recommendations

Potential Benefits

- Optimal health care delivery and symptom management
- Prolonged survival
- Enhanced quality of life

Potential Harms

- Radiation therapy side effects: erythema, sore throat, nausea
- Drug side effects

Qualifying Statements

This statement is provided as an educational service of the American Academy of Neurology (AAN). 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 American Academy of Neurology 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. The clinical context section is made available in order to place the evidence-based guideline(s) into perspective with current practice habits and challenges. No formal practice recommendations should be inferred.

Implementation of the Guideline

Description of Implementation Strategy

An implementation strategy was not provided.

Implementation Tools

Foreign Language Translations

Patient Resources

Quick Reference Guides/Physician Guides
Institute of Medicine (IOM) National Healthcare Quality Report Categories

IOM Care Need
End of Life Care
Living with Illness

IOM Domain
Effectiveness
Patient-centeredness

Identifying Information and Availability

Bibliographic Source(s)

Adaptation
Not applicable: The guideline was not adapted from another source.

Date Released
1999 Apr (revised 2009 Oct 13; reaffirmed 2013 Nov)

Guideline Developer(s)
American Academy of Neurology - Medical Specialty Society

Source(s) of Funding
American Academy of Neurology (AAN)
Guideline Committee

Quality Standards Subcommittee

Composition of Group That Authored the Guideline

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Financial Disclosures/Conflicts of Interest

Financial Disclosures

Dr. Miller serves on the editorial board of the ALS Journal; received a speaker honorarium from the AANEM; served as a consultant to Celgene, Knopp Neurosciences Inc., Teva Pharmaceutical Industries Ltd., Taiji Biomedical, Inc., Sanofi-Aventis, Novartis, and Neuraltus; and receives research support from the NIH [R01 NS 44887 (PI)] and the Muscular Dystrophy Association (PI). Dr. Jackson serves as a consultant to Knopp Neurosciences Inc. and receives research support from Knopp Neurosciences Inc., Insmed Inc., Solstice Neurosciences, Inc., the ALS Association, and NIH NINDS [U01 NS042685-0 (site PI), R01NS045087-01A2 (site PI), and N01-AR-2250 (site PI)]. Dr. Kasarskis serves as an Associate Editor for Amyotrophic Lateral Sclerosis; has received honoraria from the American Institute for Biological Studies (grant reviews); served as a consultant to Acceleron Pharma; holds equity in Amgen; and receives research support from the NIH/NINDS [R01-NS045087 (PI) and 1U01 NS049640 (site PI)]. Dr. England serves as an Associate Editor for Current Treatment Options in Neurology; received a speaker honorarium from Teva Pharmaceutical Industries Ltd.; and serves as a consultant to Talecris. Ms. Forshew has served on a scientific advisory board for the ALS Association and receives research support from the Muscular Dystrophy Association. Dr. Johnston reports no disclosures. Dr. Kalra receives research support from the ALS Association of America and the ALS Society of Canada. Dr. Katz has received research support from Pfizer Inc. Dr. Mitsumoto served on scientific advisory boards for Avanir Pharmaceuticals, Knopp Neurosciences Inc., Neuralstem, Inc., Aisai Communication Technology Co., Ltd., and Otsuka Pharmaceutical Co., Ltd.; and receives research support from Avanir Pharmaceuticals, Teva Pharmaceutical Industries Ltd., Knopp Neurosciences Inc., Sanofi-Aventis, Athena Diagnostics, Inc., Bio-Scrip, and the NIH/NINDS [DNA repository as a supplement and NIEHS Center grant]. Dr. Rosenfeld serves on the editorial board of Amyotrophic Lateral Sclerosis and has served as a consultant to Solstice Neurosciences, Inc and Aveica Group, Inc. Dr. Shoesmith receives research support from the Muscular Dystrophy Association and her spouse is employed by Biovail Pharmaceuticals Canada. Dr. Strong serves on the editorial board of Amyotrophic Lateral Sclerosis. Dr. Woolley has received research support from Pfizer Inc., Eisai Inc., and the ALS Association (co-I).

Conflict of Interest

The American Academy of Neurology is committed to producing independent, critical and truthful clinical practice guidelines (CPGs). Significant efforts are made to minimize the potential for conflicts of interest to influence the recommendations of this CPG. To the extent possible, the AAN keeps separate those who have a financial stake in the success or failure of the products appraised in the CPGs and the developers of the guidelines. Conflict of interest forms were obtained from all authors and reviewed by an oversight committee prior to project initiation. AAN limits the participation of authors with substantial conflicts of interest. The AAN forbids commercial participation in, or funding of, guideline projects. Drafts of the guideline have been reviewed by at least three AAN committees, a network of neurologists, Neurology® peer reviewers and representatives from related fields. The AAN Guideline Author Conflict of Interest Policy can be viewed at www.aan.com

Guideline Endorser(s)

American Association of Neuromuscular and Electrodiagnostic Medicine - Medical Specialty Society
Guideline Status

This is the current release of the guideline.


The American Academy of Neurology reaffirmed the currency of this guideline in 2013.

Guideline Availability

Electronic copies: A list of American Academy of Neurology (AAN) guidelines, along with a link to a Portable Document Format (PDF) file for this guideline, is available at the AAN Web site.

Print copies: Available from the AAN Member Services Center, (800) 879-1960, or from AAN, 201 Chicago Avenue South, Minneapolis, MN 55415.

Availability of Companion Documents

The following are available:


In addition, a Chinese translation of the original guideline document is available from the Neurology Journal Web site.

Patient Resources

The following is available:


Please note: This patient information is intended to provide health professionals with information to share with their patients to help them better understand their health and their diagnosed disorders. By providing access to this patient information, it is not the intention of NGC to provide specific medical advice for particular patients. Rather we urge patients and their representatives to review this material and then to consult with a licensed health professional for evaluation of treatment options suitable for them as well as for diagnosis and answers to their personal medical questions. This patient information has been derived and prepared from a guideline for health care professionals included on NGC by the authors or publishers of that original guideline. The patient information is not reviewed by NGC to establish whether or not it accurately reflects the original guideline's content.

NGC Status
This summary was completed by ECRI on February 12, 2002. The information was verified by the guideline developer as of March 29, 2002. This summary was updated by ECRI Institute on November 2, 2007, following the U.S. Food and Drug Administration advisory on Antidepressant drugs. This summary was updated by ECRI Institute on September 1, 2010. The currency of the guideline was reaffirmed by the developer in 2013 and this summary was updated by ECRI Institute on May 27, 2015. This summary was updated by ECRI Institute on June 2, 2016 following the U.S. Food and Drug Administration advisory on opioid pain medicines. This summary was updated by ECRI Institute on October 21, 2016 following the U.S. Food and Drug Administration advisory on opioid pain and cough medicines combined with benzodiazepines.

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