



Complete Summary

GUIDELINE TITLE

Practice parameter for repetitive nerve stimulation and single fiber EMG evaluation of adults with suspected myasthenia gravis or Lambert-Eaton myasthenic syndrome: summary statement.

BIBLIOGRAPHIC SOURCE(S)

AAEM Quality Assurance Committee. Practice parameter for repetitive nerve stimulation and single fiber EMG evaluation of adults with suspected myasthenia gravis or Lambert-Eaton myasthenic syndrome: summary statement. Muscle Nerve 2001 Sep; 24(9):1236-8. [36 references]

COMPLETE SUMMARY CONTENT

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- METHODOLOGY - including Rating Scheme and Cost Analysis
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SCOPE

DISEASE/CONDITION(S)

- Myasthenia gravis (MG)
- Lambert-Eaton myasthenic syndrome (LEMS)

GUIDELINE CATEGORY

Diagnosis

CLINICAL SPECIALTY

Neurology
Physical Medicine and Rehabilitation

INTENDED USERS

Physicians

GUIDELINE OBJECTIVE(S)

To present evidence-based practice parameters for the use of repetitive nerve stimulation (RNS) and single fiber electromyography (SFEMG) in the clinical diagnosis of myasthenia gravis (MG) and Lambert-Eaton myasthenic syndrome (LEMS)

TARGET POPULATION

Men and women with suspected myasthenia gravis (MG) or Lambert-Eaton myasthenic syndrome (LEMS)

INTERVENTIONS AND PRACTICES CONSIDERED

Diagnosis

1. Repetitive nerve stimulation (RNS)
2. Single fiber electromyography (SFEMG)

Note: Routine needle electromyography (EMG) and nerve conduction studies (NCSs) are considered to exclude disorders other than myasthenia gravis or Lambert-Eaton myasthenic syndrome.

MAJOR OUTCOMES CONSIDERED

Validity (sensitivity, specificity) of repetitive nerve stimulation (RNS) and single fiber electromyography (SFEMG) in confirming a clinical diagnosis of myasthenia gravis (MG) or Lambert-Eaton myasthenic syndrome (LEMS)

METHODOLOGY

METHODS USED TO COLLECT/SELECT EVIDENCE

Hand-searches of Published Literature (Primary Sources)
Hand-searches of Published Literature (Secondary Sources)
Searches of Electronic Databases

DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE

A Medline search was conducted for literature in English retrospectively through July 1998, under the Medical Subject Headings (MeSH): (1) neuromuscular junction; (2) neuromuscular transmission; (3) myasthenia gravis; (4) Lambert-Eaton; (5) myasthenic; and (6) botulism with electromyography (EMG) or nerve conduction study (NCS).

There were 545 articles identified, of which 13 articles met at least 3 of 6 criteria set previously by the American Association of Electrodiagnostic Medicine (AAEM).

An additional 21 articles were identified from review articles or the references of these first 13 articles, leading to a total of 34 articles.

NUMBER OF SOURCE DOCUMENTS

34

METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE

Weighting According to a Rating Scheme (Scheme Given)

RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE

Literature Classification Criteria

1. Prospective study.
2. Diagnosis in patient population based on clinical criteria independent of the electrodiagnostic procedure under evaluation.
3. Electrodiagnostic procedure described in sufficient detail to permit duplication of the procedure.
4. Limb temperature monitored.
5. Reference values for the electrodiagnostic test obtained with either (a) concomitant studies of a reference population, or (b) previous studies of a reference population in the same laboratory.
6. Criteria for abnormal findings clearly stated and, if the measurement is a quantitative one, the criteria for an abnormal value defined in statistically computed terms (e.g., range or mean \pm 2 standard deviations) from data derived from the reference population.

Definitions for Classification of Evidence

1. Class A evidence: studies that meet all six literature classification criteria.
2. Class B evidence: studies that meet four or five literature classification criteria.
3. Class C evidence: studies that meet three or fewer literature classification criteria.

METHODS USED TO ANALYZE THE EVIDENCE

Systematic Review

DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE

Not stated

METHODS USED TO FORMULATE THE RECOMMENDATIONS

Not stated

RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS

Definitions for Practice Recommendation Strength

1. Practice standards. Generally accepted principles for patient management that reflect a high degree of clinical certainty (Class A evidence).
2. Practice guidelines. Recommendations for patient management that reflect moderate clinical certainty (Class B evidence).
3. Practice options/advisories. Other strategies for patient management for which the clinical utility is uncertain (Class C evidence).

COST ANALYSIS

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

METHOD OF GUIDELINE VALIDATION

Peer Review

DESCRIPTION OF METHOD OF GUIDELINE VALIDATION

The American Association of Electrodiagnostic Medicine (AAEM) acknowledges Michael Venix, MD, and Donald B. Sanders, MD for reviewing the paper and making helpful suggestions.

RECOMMENDATIONS

MAJOR RECOMMENDATIONS

The definitions for the classification of evidence (Class A, B, and C) and the practice recommendation strengths (practice standards, practice guidelines, practice options/advisories) are provided at the end of the "Major Recommendations" field.

1. Guideline. Repetitive nerve stimulation (RNS) of a nerve supplying a symptomatic muscle should be performed. Abnormality in myasthenia gravis (MG) is considered to be a reproducible 10% decrement in amplitude when comparing the first stimulus to the fourth or fifth, which is found in at least one muscle. Abnormality in Lambert-Eaton myasthenic syndrome (LEMS) is considered to be a reproducible postexercise increase in amplitude of at least 100% as compared to preexercise baseline value.

The conditions recommended for RNS testing are as follows:

- a. Anticholinesterase medications withheld 12 h prior to testing, if this can be done safely.
- b. Immobilization of limb when possible.
- c. Frequency of stimulation between 2 and 5 Hz.

- d. Baseline and immediate postexercise or posttetanic 2 to 5-Hz nerve stimulation followed by stimulation at regular intervals of 30 s to 1 min, and continuing to 5 min.
 - e. Skin temperature over the recording site should be maintained as close to 35 degrees C as possible.
2. Guideline. If RNS is normal and there is high suspicion for a neuromuscular junction (NMJ) disorder, single fiber electromyography (SFEMG) of at least one symptomatic muscle should be performed. If SFEMG of one muscle is normal and clinical suspicion for a NMJ disorder is high, a second muscle should be studied.

The conditions recommended for SFEMG testing are as follows:

- a. Acceptable muscle fiber potential pairs must have an amplitude greater than 200 microvolts and a rise time less than 300 microseconds.
- b. Jitter is accurately calculated as mean consecutive difference (MCD) using the formula:

$$\text{MCD} = [IPI_1 - IPI_2] + \dots + [IPI_{n-1} - IPI_n]/n - 1$$

where IPI is the interpotential interval.

- c. A study should be considered abnormal if greater than 10% of fiber potential pairs exceed normal jitter or have impulse blockade, and/or mean jitter exceeds normal limits.
3. Option. If the patient has very mild or solely ocular symptoms and it is believed the RNS will be normal, or if the discomfort associated with RNS prevents completion of RNS, SFEMG testing may be performed in place of RNS as the initial neuromuscular junction test. In laboratories with SFEMG capability, SFEMG may be performed as the initial test for disorders of neuromuscular transmission as it is more sensitive than RNS. Routine needle electromyography (EMG) and nerve conduction studies (NCSs) may be necessary to exclude disorders other than myasthenia gravis or Lambert-Eaton myasthenic syndrome.

Definitions:

Literature Classification Criteria:

1. Prospective study.
2. Diagnosis in patient population based on clinical criteria independent of the electrodiagnostic procedure under evaluation.
3. Electrodiagnostic procedure described in sufficient detail to permit duplication of the procedure.
4. Limb temperature monitored.
5. Reference values for the electrodiagnostic test obtained with either (a) concomitant studies of a reference population, or (b) previous studies of a reference population in the same laboratory.
6. Criteria for abnormal findings clearly stated and, if the measurement is a quantitative one, the criteria for an abnormal value defined in statistically

computed terms (e.g., range or mean \pm 2 standard deviations) from data derived from the reference population.

Classification of Evidence:

1. Class A evidence: studies that meet all six literature classification criteria.
2. Class B evidence: studies that meet four or five literature classification criteria.
3. Class C evidence: studies that meet three or fewer literature classification criteria.

Practice Recommendation Strengths:

1. Practice standards. Generally accepted principles for patient management that reflect a high degree of clinical certainty (Class A evidence).
2. Practice guidelines. Recommendations for patient management that reflect moderate clinical certainty (Class B evidence).
3. Practice options/advisories. Other strategies for patient management for which the clinical utility is uncertain (Class C evidence).

CLINICAL ALGORITHM(S)

None provided

EVIDENCE SUPPORTING THE RECOMMENDATIONS

TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

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

The practice parameters are based on an extensive review of the scientific literature supporting the use of nerve conduction studies and needle electromyography in the evaluation of neuromuscular junction disorders.

BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

POTENTIAL BENEFITS

Appropriate use of repetitive nerves stimulation (RNS) and single fiber electromyography (SFEMG) for electrodiagnosis of myasthenia gravis or Lambert-Eaton myasthenic syndrome.

POTENTIAL HARMS

- The risks of electrodiagnostic testing to the patient include transient discomfort, bruise, hematoma, and infection from the needle insertion required to perform both single fiber and needle electromyography (EMG).
- The risks of electromyography to the electrodiagnosis (EDX) consultant include inadvertent needle puncture of the consultant by the needle used to

- evaluate the patient and subsequent infection by hepatitis, human immunodeficiency virus (HIV), or other communicable disease.
- The risks of repetitive nerve stimulation (RNS) include transient discomfort and accidental electric shock.

CONTRAINDICATIONS

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Contraindications to repetitive nerve stimulation (RNS) to avoid accidental electric shock include patients with pacemakers and central lines.

QUALIFYING STATEMENTS

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This statement is provided as an educational service of the American Association of Electrodiagnostic Medicine (AAEM). 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 Association of Electrodiagnostic Medicine recognizes that specific patient care decisions are the prerogative of the patient and his/her physician and are based on all of the circumstances involved.

IMPLEMENTATION OF THE GUIDELINE

DESCRIPTION OF IMPLEMENTATION STRATEGY

An implementation strategy was not provided.

INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

IOM CARE NEED

Living with Illness

IOM DOMAIN

Effectiveness
Safety

IDENTIFYING INFORMATION AND AVAILABILITY

BIBLIOGRAPHIC SOURCE(S)

AAEM Quality Assurance Committee. Practice parameter for repetitive nerve stimulation and single fiber EMG evaluation of adults with suspected myasthenia gravis or Lambert-Eaton myasthenic syndrome: summary statement. Muscle Nerve 2001 Sep; 24(9): 1236-8. [36 references]

ADAPTATION

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

DATE RELEASED

2001 Sep

GUIDELINE DEVELOPER(S)

American Association of Electrodiagnostic Medicine - Medical Specialty Society

SOURCE(S) OF FUNDING

American Association of Electrodiagnostic Medicine (AAEM)

GUIDELINE COMMITTEE

American Association of Electrodiagnostic Medicine (AAEM) Quality Assurance Committee

COMPOSITION OF GROUP THAT AUTHORED THE GUIDELINE

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FINANCIAL DISCLOSURES/CONFLICTS OF INTEREST

Not stated

GUIDELINE STATUS

This is the current release of the guideline.

An update is not in progress at this time.

GUIDELINE AVAILABILITY

Electronic copies: Not available at this time.

Print copies: Available from AAEM. To obtain an order form, please contact the Education Department at the AAEM Executive Office, 421 First Ave SW, Suite 300 E, Rochester, MN 55902; (507) 288-0100; fax, (507) 288-1225; e-mail: aaem@aaem.net. The order form is also posted on the [AAEM Web site](#).

AVAILABILITY OF COMPANION DOCUMENTS

The following is available:

- Literature review of the usefulness of nerve conduction studies and needle electromyography for the evaluation of patients with carpal tunnel syndrome. AAEM Quality Assurance Committee. Muscle Nerve. Muscle Nerve 2001 Sep; 24(9): 1239-47

Electronic copies: Not available at this time.

Print copies: Available from AAEM. To obtain an order form, please contact the Education Department at the AAEM Executive Office, 421 First Ave SW, Suite 300 E, Rochester, MN 55902; (507) 288-0100; fax, (507) 288-1225; e-mail: aaem@aaem.net. The order form is also posted on the [AAEM Web site](#).

PATIENT RESOURCES

None available

NGC STATUS

This NGC summary was completed by ECRI on October 4, 2002. The information was verified by the guideline developer on November 13, 2002.

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