

Overview

Useful For

Initial evaluation of patients aged 20 or older with symptoms and signs of acquired myasthenia gravis (MG)

Bone marrow transplant recipients with suspected graft-versus-host disease, particularly if weakness has appeared

Confirming that a recently acquired neurological disorder has an autoimmune basis (eg, MG)

Providing a quantitative baseline for future comparisons in monitoring a patient's clinical course and the response to immunomodulatory treatment

Raising likelihood of neoplasia

Profile Information

Test ID	Reporting Name	Available Separately	Always Performed
MGEAI	MG Adult Interpretation, S	No	Yes
ARBI	ACh Receptor (Muscle) Binding Ab	Yes	Yes
ARMO	ACh Receptor (Muscle) Modulating Ab	No	Yes
STR	Striational (Striated Muscle) Ab, S	Yes	Yes

Reflex Tests

Test ID	Reporting Name	Available Separately	Always Performed
GD65S	GAD65 Ab Assay, S	Yes	No
CRMWS	CRMP-5-IgG Western Blot, S	Yes	No
GANG	AChR Ganglionic Neuronal Ab, S	No	No
VGKC	Neuronal (V-G) K+ Channel Ab, S	No	No

Testing Algorithm

If acetylcholine receptor (AChR) modulating antibodies are > or =90% and striational antibodies are > or =1:120, then AChR ganglionic neuronal autoantibody, glutamic acid decarboxylase autoantibody, neuronal voltage-gated potassium channel autoantibody, and CRMP-5-IgG Western blot will be performed at an additional charge.

See [Myasthenia Gravis: Adult Diagnostic Algorithm](#) in Special Instructions.

Special Instructions

- [Myasthenia Gravis: Adult Diagnostic Algorithm](#)

Method Name

ARBI, ARMO, GANG, VGKC: Radioimmunoassay (RIA)

STR: Enzyme Immunoassay (EIA)

NY State Available

Yes

Specimen

Specimen Type

Serum

Specimen Required

Container/Tube:

Preferred: Red top

Acceptable: Serum gel

Specimen Volume:3 mL

Additional Information: Patient should have no general anesthetic or muscle-relaxant drugs in the previous 24 hours.

Forms

If not ordering electronically, complete, print, and send a [Neurology Specialty Testing Client Test Request](#) (T732) with the specimen.

Specimen Minimum Volume

2 mL

Reject Due To

Gross hemolysis	Reject
Gross lipemia	Reject
Gross icterus	Reject

Specimen Stability Information

Specimen Type	Temperature	Time	Special Container
Serum	Refrigerated (preferred)	28 days	
	Frozen	28 days	
	Ambient	72 hours	

Clinical and Interpretive

Clinical Information

Myasthenia gravis (MG) is an acquired disorder of neuromuscular transmission caused by the binding of pathogenic autoantibodies to muscle's postsynaptic nicotinic acetylcholine receptor (AChR). In a small minority of patients the pathogenic antibody is directed at the muscle-specific receptor tyrosine kinase (MuSK) antigen. The ensuing weakness in both cases reflects a critical loss of the AChR channel protein, which is required to activate the muscle action potential.

MG affects children (see MGP1 / Myasthenia Gravis [MG] Evaluation, Pediatric) as well as adults. In adults with MG there is at least a 20% occurrence of thymoma or other neoplasm. Neoplasms are an endogenous source of the antigens driving production of autoantibodies.

Autoimmune serology is indispensable for initial evaluation and monitoring of patients with acquired disorders of neuromuscular transmission. The neurological diagnosis depends on the clinical context and electromyographic findings, and is confirmed more readily by the individual patient's serological profile than by any single test.

Not all of the antibodies detected in this profile impair neuromuscular transmission (eg, antibodies directed at cytoplasmic epitopes accessible on solubilized AChR, or sarcomeric proteins that constitute the striational antigens).

If muscle acetylcholine receptor (AChR) modulating antibody value is (or exceeds) 90% AChR loss and striational antibody is detected, thymoma is likely. Reflexive testing will include collapsin response-mediated protein-5-IgG Western blot, ganglionic AChR antibody, glutamic acid decarboxylase (GAD65) antibody, and voltage-gated potassium channel complex (VGKC) antibody (which are frequent with thymoma).

Note: Single antibody tests may be requested in follow-up of patients with positive results documented in this laboratory.

See [Myasthenia Gravis: Adult Diagnostic Algorithm](#) in Special Instructions.

Reference Values

ACh RECEPTOR (MUSCLE) BINDING ANTIBODY

< or =0.02 nmol/L

ACh RECEPTOR (MUSCLE) MODULATING ANTIBODIES

0-20% (reported as ___% loss of AChR)

STRIATIONAL (STRIATED MUSCLE) ANTIBODIES

<1:120

Interpretation

The patient's autoantibody profile is more informative than the result of any single test for supporting a diagnosis of myasthenia gravis (MG), and for predicting the likelihood of thymoma (see MGT1 / Myasthenia Gravis [MG] Evaluation, Thymoma).

Muscle acetylcholine receptor (AChR) and striational autoantibodies are characteristic but not diagnostic of MG. One or both are found in 13% of patients with Lambert-Eaton Syndrome (LES), but P/Q-type calcium channel autoantibodies are very rare in MG.

Results are sometimes positive in patients with neoplasia without evidence of neurological impairment.

Titers are generally higher in patients with severe weakness, or with thymoma, but severity cannot be predicted by antibody titer.

Test results for muscle acetylcholine receptor and striational antibodies may be negative for 6 to 12 months after MG symptom onset. Only 8% of nonimmunosuppressed patients with generalized MG remain seronegative beyond 12 months for all autoantibodies in the adult MG evaluation. Of those patients 38% will have the alternative muscle-specific receptor tyrosine kinase (MuSK)-specific autoantibody.

MuSK antibody-positive patients lack thymoma, and have predominantly oculobulbar symptoms that respond to plasmapheresis and immunosuppressant therapy. They do not benefit from thymectomy.

Cautions

A positive result is not per se diagnostic of myasthenia gravis (MG). Positive values for muscle antibodies (acetylcholine receptor: AChR or striational) occur in 13% of Lambert-Eaton syndrome (LES) patients, 40% of patients with autoimmune liver disorders, approximately 10% of patients with lung cancer, and in patients with graft-versus-host disease, and recipients of D-penicillamine.

False-positive results occur most frequently in the bioassay for AChR modulating antibody; serum redraw will be requested when only this assay yields a positive result. Curare-like drugs used during general anesthesia can yield transient false-positive results for AChR modulating antibodies.

Seropositive rates differ in different laboratories.

This test should not be requested in patients who have recently received radioisotopes, therapeutically or diagnostically, because of potential assay interference. The specific waiting period before specimen collection will depend on the isotope administered, the dose given and the clearance rate in the individual patient. Specimens will be screened for radioactivity prior to analysis. Radioactive specimen received in the laboratory will be held 1 week and assayed if sufficiently decayed, or canceled if radioactivity remains.

The presence of alpha-bungarotoxin antibodies may interfere with the ACh receptor (muscle) binding antibody assay.

Clinical Reference

1. Lennon VA: Serological profile of myasthenia gravis and distinction from the Lambert-Eaton myasthenic syndrome. *Neurology* 1997;48(Suppl 5):S23-S27
2. Harper CM, Lennon VA: Lambert-Eaton syndrome. In *Current Clinical Neurology: Myasthenia Gravis and Related Disorders*. Edited by HJ Kaminski. Totowa, NJ, Humana Press, 2002, pp 269-291
3. Hoch W, McConville J, Helms S, et al: Auto-antibodies to the receptor tyrosine kinase MuSK in patients with myasthenia gravis without acetylcholine receptor antibodies. *Nat Med* 2001 Mar;7(3):365-368
4. Chan KH, Lachance DH, Harper CM, Lennon VA: Frequency of seronegativity in adult-acquired generalized myasthenia gravis. *Muscle Nerve* 2007;36:651-658
5. Skjei KL, Lennon VA, Kuntz NL: Muscle specific kinase antibody-associated myasthenia gravis in children; clinical, serologic, electrophysiologic and pathologic characteristics and response to treatment. *Ann Neurol*

62(S11):S145-S146

Performance

Method Description

Muscle acetylcholine receptor (AChR) binding (IgG and IgM) are measured quantitatively by immunoprecipitation assays. Fetal and adult, detergent-solubilized, acetylcholine receptors (extracted from cultures of rhabdomyosarcoma [RD] cells) labeled with (125)I-alpha-bungarotoxin are incubated with patient serum. AChR modulating antibody is detected in a bioassay; (125)I-bungarotoxin measures percent loss of AChR from viable, noninnervated, monolayer cultures of human muscle cells following 14-hour incubation with patient's serum. The EIA used to detect striational antibodies (IgG, IgM, and IgA) employs as antigen a mixture of sarcomeric proteins extracted from healthy adult rat skeletal muscle. (Griesmann GE, Kryzer TJ, Lennon VA: Autoantibody profiles of myasthenia gravis and Lambert-Eaton myasthenic syndrome. In Manual of Clinical and Laboratory Immunology. Sixth edition. Edited by NR Rose, RG Hamilton, B Detrick. Washington, DC, ASM Press, 2002, pp 1005-1012)

PDF Report

No

Day(s) and Time(s) Test Performed

ACh receptor (muscle) binding antibody:

Monday through Friday; 11 a.m., 6 p.m., 10 p.m.

Saturday; 6 a.m.

Sunday; 6 a.m., 10 a.m.

ACh receptor (muscle) modulating antibodies:

Monday through Thursday; 2 p.m.

Saturday; 8 a.m.

Striational (striated muscle) antibodies:

Monday through Friday; 4 a.m., 3 p.m.

Saturday; 6 a.m.

CRMP-5-IgG Western blot:

Monday, Wednesday, Friday; 8 a.m.

AChR ganglionic neuronal antibody:

Monday through Friday; 11 a.m., 6 p.m.

Saturday; 6 a.m.

Sunday; 6 a.m.

Neuronal VGKC autoantibody:

Monday through Friday; 11 a.m., 6 p.m.

Saturday; 6 a.m.

Sunday; 6 a.m.

GAD65 antibody assay:

Monday through Friday; 6 a.m., 4 p.m.

Analytic Time

3 days

Maximum Laboratory Time

7 days

Specimen Retention Time

28 days

Performing Laboratory Location

Rochester

Fees and Codes**Fees**

- Authorized users can sign in to [Test Prices](#) for detailed fee information.
- Clients without access to Test Prices can contact [Customer Service](#) 24 hours a day, seven days a week.
- Prospective clients should contact their Regional Manager. For assistance, contact [Customer Service](#).

Test Classification

This test was developed and its performance characteristics determined by Mayo Clinic in a manner consistent with CLIA requirements. This test has not been cleared or approved by the U.S. Food and Drug Administration.

CPT Code Information

83519 x 2

83520

83519 x 2 (if appropriate)

84182 (if appropriate)

86341 (if appropriate)

LOINC® Information

Test ID	Test Order Name	Order LOINC Value
MGA1	MG Evaluation, Adult	53706-8

Result ID	Test Result Name	Result LOINC Value
34272	MG Adult Interpretation, S	69048-7
8338	ACh Receptor (Muscle) Binding Ab	11034-6
8879	ACh Receptor (Muscle) Modulating Ab	30192-9
8746	Striational (Striated Muscle) Ab, S	94817-4