

## Overview

### Useful For

Recommended for initial investigation of patients presenting at less than age 20 with a defect of neuromuscular transmission

Confirming that a recently acquired neurological disorder has an autoimmune basis

Distinguishing acquired myasthenia gravis from congenital myasthenic syndromes (persistently seronegative)

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

### Profile Information

Test ID	Reporting Name	Available Separately	Always Performed
MGEPI	MG Pediatric Interpretation, S	No	Yes
ARBI	ACh Receptor (Muscle) Binding Ab	Yes	Yes
ARMO	ACh Receptor (Muscle) Modulating Ab	No	Yes

### Testing Algorithm

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

### Special Instructions

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

### Method Name

Radioimmunoassay (RIA)

### NY State Available

Yes

## Specimen

### Specimen Type

Serum

### Specimen Required

#### Container/Tube:

**Preferred:** Red top

**Acceptable:** Serum gel

**Specimen Volume:** 2 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

1.5 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 about 3% of cases the pathogenic antibody is directed at the functionally associated muscle-specific receptor tyrosine kinase (MuSK). The outcome is a critical loss of the AChR channel protein, which is required to activate the muscle action potential.

Amongst North American Caucasian children (ie, aged 1-18), MG affects prepubertal boys and girls with equal frequency. Spontaneous remissions are relatively frequent. Females predominate (4.5:1) after puberty. Amongst black children with MG, females predominate (2:1) in all age groups, and remissions are infrequent, regardless of therapy.

Congenital MG is a hereditary nonautoimmune disorder characterized by defects in AChR or other synaptic proteins.

Autoimmune serology is indispensable for both initial evaluation and monitoring the course of patients with acquired disorders of neuromuscular transmission. The neurological diagnosis depends on the clinical context, electromyographic findings, and response to anticholinesterase administration. MG is confirmed more readily by a serological profile than by any single test.

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

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See [Myasthenia Gravis: Pediatric 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)

## Interpretation

Muscle acetylcholine receptor (AChR) autoantibodies are characteristic but not diagnostic of myasthenia gravis (MG). They are found in 13% of patients with Lambert-Eaton Syndrome (LES), which is rare in children. The patient's autoantibody profile is more informative than the result of any single test for supporting a diagnosis of MG.

Titers of AChR antibodies are generally higher in patients with severe weakness, but severity cannot be predicted by antibody titer. Seronegativity is more frequent in children with prepubertal onset of acquired MG (33%-50%) than in adults (<10%). Thymoma is rare under age 20, and striational antibodies (see STR / Striational [Striated Muscle] Antibodies, Serum) also are rare, except in the context of MG related to neoplasia (usually thymoma or neuroblastoma), graft-versus-host disease, autoimmune liver disease, or D-penicillamine therapy. This laboratory has recently noted muscle-specific receptor tyrosine kinase antibody in children with "seronegative" acquired MG, but the frequency of this antibody in pediatric MG has not been determined.

## Cautions

A positive result in this evaluation is not per se diagnostic of myasthenia gravis (MG). Positive values for muscle acetylcholine receptor (AChR) antibodies occur in 10% of Lambert-Eaton syndrome patients, in children with graft-versus-host disease, and recipients of D-penicillamine (with and without clinically evident MG), and in children with paraneoplastic neurological disorders related to neuroblastoma, thymoma, and chondroblastoma (ie, seropositivity is not restricted to MG). Children with autoimmune liver disorders may be anticipated, like adults, to have unexplained AChR or striational antibodies (data not available).

Seronegativity does not exclude the diagnosis of autoimmune MG.

A minority of patients lacking detectable AChR antibodies have the recently discovered muscle-specific receptor tyrosine kinase antibodies.

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. AChR blocking antibody is the least frequently encountered AChR antibody specificity, and is never positive with a negative AChR modulating value. Curare-like drugs used during general anesthesia can yield a false-positive AChR blocking antibody result.

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 specimens 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.

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**Clinical Reference**

1. Andrews PI, Massey JM, Howard JF Jr, Sanders DB: Race, sex, and puberty influence onset, severity, and outcome in juvenile myasthenia gravis. *Neurology* 1994 July;44(7):1208-1214
2. Lennon VA: Serological profile of myasthenia gravis and distinction from the Lambert-Eaton myasthenic syndrome. *Neurology* 1997;48(Suppl 5):S23-S27
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. Joshi DD, Anderson PM, Matsumoto J, et al: Metastatic chondroblastoma with elevated creatine kinase and paraneoplastic neurologic autoimmunity. *J Pediatr Hematol Oncol* 2003;25:900-904

**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 antibodies 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. (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.

**Analytic Time**

3 days

**Maximum Laboratory Time**

5 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**

See Individual Test IDs

**CPT Code Information**

83519 x 2

**LOINC® Information**

Test ID	Test Order Name	Order LOINC Value
MGP1	MG Eval, Pediatric	53705-0

Result ID	Test Result Name	Result LOINC Value
8338	ACh Receptor (Muscle) Binding Ab	11034-6
8879	ACh Receptor (Muscle) Modulating Ab	30192-9
34275	MG Pediatric Interpretation, S	69048-7