

Test Definition: AAGTS

Alpha-3 Beta-4 Ganglionic Neuronal
Acetylcholine Receptor Binding Antibody, Flow
Cytometry Titer, Serum (Non-Orderable)

Overview

Useful For

Only orderable as a reflex. For more information see:
DYS2 / Dysautonomia, Autoimmune/Paraneoplastic Evaluation, Serum
GID2 / Gastrointestinal Dysmotility, Autoimmune/Paraneoplastic Evaluation, Serum

Evaluating patients with suspected autoimmune autonomic ganglionopathy and autoimmune gastrointestinal dysmotility using serum specimens

Reporting alpha-3 beta-4 ganglionic neuronal acetylcholine receptor binding antibody end titer results of as part of a profile

Method Name

Only orderable as a reflex. For more information see:
-DYS2 / Dysautonomia, Autoimmune/Paraneoplastic Evaluation, Serum
-GID2 / Gastrointestinal Dysmotility, Autoimmune/Paraneoplastic Evaluation, Serum

Flow Cytometry

NY State Available

Yes

Specimen

Specimen Type

Serum

Specimen Required

Only orderable as a reflex. For more information see:
-DYS2 / Dysautonomia, Autoimmune/Paraneoplastic Evaluation, Serum
-GID2 / Gastrointestinal Dysmotility, Autoimmune/Paraneoplastic Evaluation, Serum

Specimen Minimum Volume

Serum: 1 mL

Reject Due To

Gross	Reject
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hemolysis	
Gross lipemia	Reject
Gross icterus	Reject

Specimen Stability Information

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

Clinical & Interpretive

Clinical Information

The assay for ganglionic acetylcholine receptor binding antibodies (alpha-3 beta-4) is most useful as a screening test for autoimmune autonomic ganglionopathy (AAG), a form of autoimmune dysautonomia which can manifest as pandysautonomia (several of fixed pupillary dilatation, orthostatic hypotension, sweat loss, gastroparesis, small bowel pseudo-obstruction, constipation and urinary retention due to bladder hypocontractility) or limited forms such as autoimmune gastrointestinal dysmotility. Occasionally, AAG may occur in a paraneoplastic context accompanying diverse carcinomas or thymoma. AAG is generally an immune therapy responsive disease.

Reference Values

Only orderable as a reflex. For more information see:
-DYS2 / Dysautonomia, Autoimmune/Paraneoplastic Evaluation, Serum
-GID2 / Gastrointestinal Dysmotility, Autoimmune/Paraneoplastic Evaluation, Serum

<1:10

Interpretation

Seropositivity for ganglionic acetylcholine receptor binding autoantibodies supports the clinical diagnosis of autoimmune autonomic failure with neurological presentations including orthostatic hypotension, constipation, dilated pupils, dry eyes and dry mouth, fainting, urinary retention. Immune therapies such as corticosteroids, plasma exchange and intravenous immunoglobulin may lead to disease remission.

Cautions

Negative results do not exclude the diagnosis of autoimmune autonomic failure.

Clinical Reference

1. Vernino S, Auger RG, Emslie-Smith AM, Harper CM, Lennon VA. Myasthenia, thymoma, presynaptic antibodies, and a continuum of neuromuscular hyperexcitability. *Neurology*. 1999;53(6):1233-1239

2. McKeon A, Lennon VA, Lachance DH, Fealey RD, Pittock SJ. Ganglionic acetylcholine receptor autoantibody: oncological, neurological, and serological accompaniments. *Arch Neurol*. 2009;66(6):735-741.

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doi:10.1001/archneurol.2009.78

3. Cutsforth-Gregory JK, McKeon A, Coon EA, et al. Ganglionic antibody level as a predictor of severity of autonomic failure. Mayo Clin Proc. 2018;93(10):1440-1447

4. Karagiorgou K, Dandoulaki M, Mantegazza R, et al. Novel Cell-Based Assay for Alpha-3 Nicotinic Receptor Antibodies Detects Antibodies Exclusively in Autoimmune Autonomic Ganglionopathy. J Neurol Neuroimmunol Neuroinflamm. 2022;9(3):e1162

Performance

Method Description

This assay is a live cell-based binding assay using flow cytometry to detect ganglionic acetylcholine receptor (AChR) binding antibodies in patient serum. Human embryonic kidney cells (HEK293) transiently expressing human alpha-3 beta-4 ganglionic AChR subunits, along with accessory proteins required for receptor expression, are used as the antigen source. Receptor expression is enhanced prior to analysis under controlled culture conditions. Cells are incubated with diluted patient serum to allow binding of AChR-specific antibodies to surface-expressed receptors. Bound human IgG is detected using a fluorophore-conjugated anti-human IgG secondary antibody, with a viability dye included to exclude non-viable cells. Fluorescence intensity is measured by flow cytometry, and antibody binding is assessed by comparing signal from AChR-expressing cells to control cells lacking receptor expression. A humanized AChR-specific monoclonal antibody is used as an assay calibrator. The IgG binding index (IBI) of patient samples is compared to the assay calibrator to calculate a cut-off Index (COI). Patient samples with a COI greater than or equal to 1.0 are reported as positive. Positive samples are further titrated to determine the end-point dilution, defined as the highest serum dilution that remains above the calibrator.(Unpublished Mayo method)

PDF Report

No

Day(s) Performed

Varies

Report Available

3 to 7 days

Specimen Retention Time

2 days

Performing Laboratory Location

Mayo Clinic Laboratories - Rochester Main Campus

Fees & Codes

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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 account representative. 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. It has not been cleared or approved by the US Food and Drug Administration.

CPT Code Information

86256

LOINC® Information

Test ID	Test Order Name	Order LOINC® Value
AAGTS	AChR Alpha3 Gang Flow Titer, S	In Process

Result ID	Test Result Name	Result LOINC® Value
623083	AChR Alpha3 Gang Flow Titer, S	In Process