

Overview**Useful For**

Supporting the diagnosis of IgG4-related disease

Method Name

Nephelometry

NY State Available

Yes

Specimen**Specimen Type**

Serum

Ordering Guidance

This test only quantitates the IgG4 protein. If quantitation of all IgG subclass types is wanted, order IGGS / IgG Subclasses, Serum.

Specimen Required

Patient Preparation: Fasting preferred but not required

Container/Tube:

Preferred: Serum gel

Acceptable: Red top

Specimen Volume: 1 mL

Specimen Minimum Volume

0.5 mL

Reject Due To

Gross hemolysis	OK
Gross lipemia	Reject
Gross icterus	OK

Specimen Stability Information

Specimen Type	Temperature	Time	Special Container
Serum	Refrigerated (preferred)	14 days	
	Ambient	14 days	
	Frozen	14 days	

Clinical and Interpretive

Clinical Information

The most abundant immunoglobulin (Ig) isotype in human serum is immunoglobulin G (IgG). IgG immunoglobulins are comprised of 4 subclasses designated IgG1 through IgG4. Of total IgG, approximately 65% is IgG1, 25% is IgG2, 6% is IgG3, and 4% is IgG4. Each IgG subclass contains structurally unique portions of the constant region of the gamma heavy chain.

IgG subclass 4-related disease is a recently recognized syndrome of unknown etiology most often occurring in middle-aged and older men. Several organ systems can be involved and encompasses many previous and newly described diseases such as type1 autoimmune pancreatitis; Mikulicz disease and sclerosing sialadenitis; inflammatory orbital pseudotumor; chronic sclerosing aortitis; Riedel thyroiditis, a subset of Hashimoto thyroiditis; IgG4-related interstitial pneumonitis; and IgG4-related tubulointerstitial nephritis. Each of these entities is characterized by tumor-like swelling of the involved organs with infiltrative, predominately IgG4-positive, plasma cells with accompanying "storiform" fibrosis. In addition, elevated serum concentrations of IgG4 are found in 60% to 70% of patients diagnosed with IgG4-related disease.

The diagnosis of IgG4-related disease requires a tissue biopsy of the affected organ demonstrating the aforementioned histological features. It is recommended that patients suspected of having an IgG4-related disease have their serum IgG4 level measured.

Reference Values

0-<5 months: < or =19.8 mg/dL

5-<9 months: < or =20.8 mg/dL

9-<15 months: < or =22.0 mg/dL

15-<24 months: < or =23.0 mg/dL

2-<4 years: 0.4-49.1 mg/dL

4-<7 years: 0.8-81.9 mg/dL

7-<10 years: 1.0-108.7 mg/dL

10-<13 years: 1.0-121.9 mg/dL

13-<16 years: 0.7-121.7 mg/dL

16-<18 years: 0.3-111.0 mg/dL

> or =18 years: 2.4-121.0 mg/dL

Interpretation

Elevated levels of IgG4 are consistent with, but not diagnostic of, IgG4-related disease.

Cautions

Elevations in serum IgG4 concentrations are not specific to IgG4-related disease; they are also found in disorders

such as multicentric Castleman disease, allergic disorders, Churg-Strauss syndrome, sarcoidosis, and a large number of other conditions.

Clinical Reference

1. Cheuk W, Chan JK: IgG4-related sclerosing disease: a critical appraisal of an evolving clinicopathologic entity. *Adv Anat Pathol* 2010;17(5):303-332
2. Zen Y, Nakanuma Y: IgG4-related disease: a cross-sectional study of 114 cases. *Am J Surg Pathol* 2010;34(12):1812-1819
3. Bateman AC, Deheragoda MG: IgG4-related systemic sclerosing disease-an emerging and under-diagnosed condition. *Histopathology* 2009;55(4):373-383

Performance**Method Description**

In this Siemens Nephelometer II method, the light scattered onto the antigen-antibody complexes is measured. The intensity of the measured scattered light is proportional to the amount of antigen-antibody complexes in the sample under certain conditions. If the antibody volume is kept constant, the signal behaves proportionally to the antigen volume.

A reference curve is generated by a standard with a known antigen content on which the scattered light signals of the samples can be evaluated and calculated as an antigen concentration. Antigen-antibody complexes are formed when a sample containing antigen and the corresponding antiserum are put into a cuvette. A light beam is generated with an LED, which is transmitted through the cuvette. The light is scattered onto the immuno-complexes that are present. Antigen and antibody are mixed in the initial measurement, but no complex is formed yet. An antigen-antibody complex is formed in the final measurement.

The result is calculated by subtracting value of the final measurement from the initial measurement. The distribution of intensity of the scattered light depends on the ratio of the particle size of the antigen-antibody complexes to the radiated wavelength. (Unpublished Mayo method; Instruction manual: Siemens Nephelometer II, Version 3, Siemens, Inc., Newark, DE, 2008)

PDF Report

No

Day(s) Performed

Monday through Friday

Report Available

1 to 2 days

Specimen Retention Time

14 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 has been cleared, approved, or is exempt by the US Food and Drug Administration and is used per manufacturer's instructions. Performance characteristics were verified by Mayo Clinic in a manner consistent with CLIA requirements.

CPT Code Information

82787

LOINC® Information

Test ID	Test Order Name	Order LOINC Value
IGGS4	Immunoglobulin Subclass IgG4, S	2469-5

Result ID	Test Result Name	Result LOINC Value
IGGS4	Immunoglobulin Subclass IgG4, S	2469-5