Test Definition: IGGS4

IgG4, Immunoglobulin Subclasses, Serum

Overview

Useful For
Supporting the diagnosis of IgG4-related disease

Method Name
Turbidimetry

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 desired, order IGGS / IgG Subclasses, Serum.

Specimen Required
Patient Preparation:
Fasting: 12 hours, preferred but not required
Supplies: Sarstedt Aliquot Tube, 5 mL (T914)
Collection Container/Tube:
Preferred: Serum gel
Acceptable: Red top
Submission Container/Tube: Plastic vial
Specimen Volume: 1 mL serum
Collection Instructions: Centrifuge and aliquot serum into a plastic vial.

Specimen Minimum Volume
Serum: 0.5 mL

Reject Due To

Gross hemolysis	Reject
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 & Interpretive

Clinical Information

The most abundant immunoglobulin isotype in human serum is 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 systemic inflammatory disease of unknown etiology, most often occurring in middle-aged and older men. Several organ systems can be involved, and the disease encompasses many previous and newly described diseases such as 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. These entities may be characterized by tumor-like swelling of the involved organs with infiltration by numerous IgG4-positive plasma cells with accompanying fibrosis. In addition, elevated serum concentrations of IgG4 are found in at least 50% of patients diagnosed with IgG4-related disease.

The diagnosis of IgG4-related disease may require 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 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: < or =49.1 mg/dL
4-<7 years: < or =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: < or =121.7 mg/dL
16-<18 years: < or =111.0 mg/dL
> or =18 years: 2.4-121.0 mg/dL

Interpretation

Elevated concentration of IgG4 is 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 other conditions.

Undetected antigen excess is a rare event but cannot be excluded. Results should always be interpreted in conjunction with other laboratory tests and clinical evidence.

Clinical Reference

1. Cheuk W, Chan JKC. 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
4. Vidarsson G, Dekkers G, Rispens T. IgG subclasses and allotypes: from structure to effector functions. Front Immunol. 2014;5:520
5. Maslinska M, Dmowska-Chalaba J, Jakubaszek M. The role of IgG4 in autoimmunity and rheumatic diseases. Front Immunol. 2022;12:787422
6. Wallace ZS, Naden RP, Chari S, et al. The 2019 American College of Rheumatology/European League Against Rheumatism classification criteria for IgG4-related disease. Ann Rheum Dis. 2020;79(1):77-87. doi:10.1136/annrheumdis-2019-216561

Performance

Method Description

The determination of the soluble antigen concentration by turbidimetric methods involves the reaction with specific antiserum to form insoluble complexes. When light is passed through the suspension formed a portion of the light is transmitted and focused onto a photodiode by an optical lens system. The amount of transmitted light is indirectly proportional to the specific protein concentration in the test sample.

Concentrations are automatically calculated by reference to a calibration curve stored within the instrument.(Package inserts: Optilite IgG4. The Binding Site Group, Ltd; 07/2019)

PDF Report

No

Day(s) Performed

Monday through Friday

Report Available

1 to 2 days

Specimen Retention Time

14 days

Performing Laboratory Location

Fees & 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 account representative. 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	IgG4, Ig Subclasses	2469-5

Result ID	Test Result Name	Result LOINC® Value
IGGS4	IgG4, Ig Subclasses	2469-5