

Overview

Useful For

Investigation of immune deficiency due to IgA2 deficiency

Evaluating patients with anaphylactic transfusion reactions

Method Name

Turbidimetry

NY State Available

Yes

Specimen

Specimen Type

Serum

Specimen 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

Collection Instructions: Centrifuge and aliquot serum into a plastic vial.

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 | Ambient | 7 days | |

| | | | |
|--|--------------------------|--------|--|
| | Refrigerated (preferred) | 7 days | |
| | Frozen | 7 days | |

Clinical & Interpretive

Clinical Information

Immunoglobulin A (IgA), the predominant immunoglobulin secreted at mucosal surfaces and the second most abundant immunoglobulin in serum, consists of 2 subclasses, IgA1 and IgA2. These subclasses differ in their molecular structure and tissue distribution. IgA1 is the major (approximately 80%) subclass in serum. It has a longer hinge region making it more susceptible to proteolytic cleavage. IgA2 is the major subclass in secretions such as milk, tears, sweat, and saliva. Although IgA deficiency is a common defect (1 in 700), it is usually asymptomatic. IgA deficiency with or without IgG subclass deficiency, however, can lead to recurrent pulmonary and gastrointestinal infections. In selective IgA deficiency, both IgA1 and IgA2 are deficient. It is also possible that only one of the subclasses is deficient. Some infections (eg, recurrent sinopulmonary infections with *Haemophilus influenzae*) may be related to a deficiency of IgA2, even in the presence of normal total IgA concentrations.

Paradoxically, bacterial infections may also cause IgA deficiency. IgA2 is more resistant to bacterial destruction than IgA1. Certain bacteria can cleave and inactivate IgA1, but not IgA2, thus depleting most of the IgA. In the presence of a concurrent IgA2 deficiency, infection by these organisms results in an apparent IgA deficiency.

IgA deficiency is a cause of anaphylactic transfusion reactions. In these situations, patients who are IgA deficient produce anti-IgA antibodies that react with IgA present in the transfusion product. While transfusion reactions typically occur in patients who have no detectable concentrations of IgA, they can also occur in patients with measurable IgA. In these situations, the complete deficiency of 1 of the IgA subclasses may be the cause of the transfusion reactions.

Reference Values

IgA

1-3.9 years: 5-194 mg/dL

4-6.9 years: 16-210 mg/dL

7-9.9 years: 27-227 mg/dL

10-11.9 years: 35-241 mg/dL

12-13.9 years: 43-252 mg/dL

14-15.9 years: 50-263 mg/dL

16-17.9 years: 57-274 mg/dL

>18 years: 85-499 mg/dL

IgA1

1-3.9 years: 6-163 mg/dL

4-6.9 years: 16-186 mg/dL

7-9.9 years: 26-209 mg/dL

10-11.9 years: 34-228 mg/dL

12-13.9 years: 40-243 mg/dL

14-15.9 years: 46-259 mg/dL

16-17.9 years: 53-274 mg/dL

>18 years: 76-328 mg/dL

IgA2

1-3.9 years: <0.5-12.4 mg/dL

4-6.9 years: <0.5-25.7 mg/dL

7-9.9 years: 1.5-38.9 mg/dL

10-11.9 years: 2.9-49.9 mg/dL

12-13.9 years: 4.0-58.7 mg/dL

14-15.9 years: 5.2-67.5 mg/dL

16-17.9 years: 6.3-76.3 mg/dL

>18 years: 6.9-114.3 mg/dL

Interpretation

Low concentrations of IgA2 with normal amounts of IgA1 suggest an IgA2 deficiency.

Elevated concentrations of IgA2 with normal or low amounts of IgA1 suggest a clonal plasma cell proliferative disorder secreting a monoclonal IgA2.

Increased total IgA concentrations may also be seen in benign disorders (eg, infection, inflammation, allergy), hyper IgD syndrome with periodic fever, and monoclonal gammopathies (eg, myeloma, monoclonal gammopathies of undetermined significance [MGUS]).

Cautions

Quantitation of specific proteins by nephelometric means may not be possible in lipemic sera due to the extreme light scattering properties of the specimen. Turbidity and particles in the specimen may result in extraneous light scattering signals, resulting in variable specimen analysis.

Clinical Reference

- Schauer U, Stemberg F, Rieger CHL, et al. Establishment of age-dependent reference values for IgA subclasses. *Clin Chim Acta*. 2003;328(1-2):129-133
- Saulsbury FT. Hyperimmunoglobulinemia D and periodic fever syndrome (HIDS) in a child with normal serum IgD, but increased serum IgA concentration. *J Pediatrics*. 2003;143(1):127-129
- Popovsky MA. *Transfusion Reactions*. American Association of Blood Banks, 3rd ed, 2007
- Derksen VFAM, Allaart CF, Van der Helm-Van Mil AHM, Huizinga TWJ, Toes REM, van der Woude D. In rheumatoid arthritis patients, total IgA1 and IgA2 levels are elevated: implications for the mucosal origin hypothesis. *Rheumatology (Oxford)*. 2022;62(1):407-416. doi:10.1093/rheumatology/keac237
- Dietzen DJ, Willrich MAV. Amino acids, peptides, and proteins. In: Rifai N, Chiu RWK, Young I, Burnham CAD, Wittwer CT, eds. *Tietz Textbook of Laboratory Medicine*. 7th ed. 2023:chap 31
- Steffen U, Koeleman CA, Sokolova MV, et al. IgA subclasses have different effector functions associated with distinct glycosylation profiles. *Nat Commun*. 2020;11(1):120. Published 2020 Jan 8. doi:10.1038/s41467-019-13992-8

Performance**Method Description**

The determination of 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 IgA Kit. The Binding Site Group, Ltd; ver 5, 08/2015; Optilite IgA1. The Binding Site Group, Ltd; ver 19, 04/2016; Optilite IgA2. The Binding Site Group, Ltd; ver 11, 02/2015)

PDF Report

No

Day(s) Performed

Tuesday, Friday

Report Available

Same day/1 to 3 days

Specimen Retention Time

14 days

Performing Laboratory Location

Rochester

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

82784

82787 x 2

LOINC® Information

| Test ID | Test Order Name | Order LOINC® Value |
|---------|-------------------|--------------------|
| IGAS | IgA Subclasses, S | 87552-6 |

| Result ID | Test Result Name | Result LOINC® Value |
|-----------|------------------|---------------------|
|-----------|------------------|---------------------|

| | | |
|-------|------|--------|
| IGA_ | IgA | 2458-8 |
| IGA1_ | IgA1 | 6886-6 |
| IGA2_ | IgA2 | 6939-3 |