Pemphigus Antibodies, IgA by IIF

ARUP LABORATORIES

Patient: [redacted]
DOB: [redacted] Age: [redacted] Sex: [redacted]
Patient Identifiers: [redacted]

Visit Number (FIN): [redacted]

Client: ARUP Example Report Only
500 Chipeta Way
Salt Lake City, UT 84108

Physician: TEST TEST

ARUP Test Code: 0092106

Collection Date: 11/03/2023
Received in lab: 11/06/2023
Completion Date: 11/15/2023

Immunodermatology Serum Test Report Navigation Guide

The Immunodermatology TESTING REPORT from the University of Utah follows "See Note" and is arranged as outlined below on the following pages:

CLINICAL INFORMATION
This content is provided by the ordering clinician and includes the reason for testing.

Specimen Details
This includes specimen identification with collected and received dates.

DIAGNOSTIC INTERPRETATION
This is a synopsis of key findings from the testing and their diagnostic relevance.

RESULTS
This section reports the discrete finding and value of each test component, along with the reference range.

COMMENTS
Specific

These comments provide an explanation of the test results as they relate to clinical considerations, and include reference to any concurrent and/or previous testing.

General

These comments summarize fundamental information about the test(s) and the component(s) assessed to aid in interpretation of their clinical applicability.

TESTING METHODS
The section lists the procedures performed, the test source(s), and the applicable laboratory developed test disclaimer(s).

TEST RESULTS SUMMARY CHART
A chart tabulating results of tests ordered for the patient by the same client is included if previous and/or concurrent testing has been performed.

ELISA RESULTS GRAPH
A graph of ELISA results also is included if previous and/or concurrent testing has been performed; the graph may be found on a subsequent page.

For testing algorithm and additional information, refer to:
arupconsult.com/content/immunobullous-skin-diseases-screening

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Chart continues on following page(s)

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IMMUNODERMATOLOGY LABORATORY REPORT

[redacted]

Submitter

ARUP Sendouts

Pemphigus Antibodies, IgA by IIF (Final result)

TESTING REPORT follows "See Note"
See Note

CLINICAL INFORMATION
Blisters and pustules with pruritus. Presumptive diagnosis is dermatitis herpetiformis, folliculitis, IgA pemphigus.

Specimen Details
[redacted] ; Collected: 11/3/2023; Received: 11/6/2023

DIAGNOSTIC INTERPRETATION

Positive IgA cell surface/intercellular substance antibodies by indirect immunofluorescence, supporting the diagnosis of IgA pemphigus

(See Results and Comments including further testing considerations)

RESULTS
Indirect Immunofluorescence (IIF)

Cell Surface (CS)/Intercellular Substance (ICS) IgA Antibodies		
IgA:	Positive, titer 1:640 (H), monkey esophagus substrate	
	Positive, titer 1:160 (H), intact human skin	

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substrate

Reference Range:
Negative - Titer less than 1:10
Borderline - Titer 1:10
Positive (H) - Titer greater than 1:10

(H) = high/positive

COMMENTS

Specific

These indirect immunofluorescence testing results, demonstrating positive IgA cell surface (CS), also known as intercellular substance (ICS), serum antibodies reacting with both monkey esophagus substrate and intact human skin substrate, support the diagnosis of IgA pemphigus. IgA pemphigus is a rare type of pemphigus, also known as intercellular IgA dermatosis. IgA pemphigus presents as two major subtypes, the subcorneal pustular dermatosis (SPD) type and the intraepidermal neutrophilic (IEN) type; however, three other IgA pemphigus variants are recognized, IgA-pemphigus vegetans, IgA-pemphigus vulgaris, and unclassified IgA pemphigus. Moreover, IgA CS/ICS antibodies may be expressed in some pemphigus variants along with IgG CS/ICS antibodies.

Clinical correlation is needed, including with direct immunofluorescence findings on a biopsy specimen and treatment status, with further clinical evaluation as indicated. Additional serum testing for IgG CS/ICS antibodies by indirect immunofluorescence and ELISAs may be performed on this serum specimen by contacting ARUP Client Services at 1-800-242-2787, option 2, with add-on test request for: Pemphigus Antibody Panel, IgG (ARUP test number 0090650).

Detection, levels, and patterns of diagnostic antibodies may fluctuate with disease manifestations. Monitoring serum antibody profiles by indirect immunofluorescence and antibody levels by ELISAs may aid in assessing disease expression and activity, including response to therapy.

If it would be helpful to discuss the patient case with this report, contact ARUP Client Services at 1-800-242-2787, option 2, and ask to speak with the Immunodermatology Laboratory at the University of Utah regarding patient results.

General

IgA cell surface (CS) antibodies, also known as intercellular substance (ICS) antibodies, are positive in patients with IgA pemphigus and in some pemphigus variants along with positive IgG CS/ICS antibodies (References). IgA CS/ICS antibodies are typically not detected in normal

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individuals, in patients with other immunobullous diseases, and in individual patients whose IgA pemphigus is minimal and/or under therapeutic control, although cell surface reactivity may be observed transiently and/or nonspecifically in normal individuals and in patients with drug reactions, infections, and other mucocutaneous diseases. Monkey esophagus and intact normal human skin substrates may demonstrate differing sensitivities and specificities for disease-associated antibodies and, when tested together, increase the likelihood of detecting IgA CS/ICS antibodies.

Positive IgA CS/ICS antibody reactivity can be:
- Consistent with IgA pemphigus variants, including evolving or treated disease;
- Co-expressed with IgG CS/ICS antibodies in pemphigus foliaceus and pemphigus vulgaris;
- Observed in non-classical forms of pemphigus, including pemphigus herpetiformis, paraneoplastic pemphigus, and intercellular IgG/IgA dermatosis; and
- Found transiently and/or nonspecifically in normal individuals and in patients with various mucocutaneous disorders including drug reactions and infections.

Approximately 40 percent of patients with nonclassical IgG/IgA pemphigus have an underlying systemic disease when diagnosed, malignancy being the most common.

References:
- Mentink LF, de Jong MC, Kloosterhuis GJ, et al. Coexistence of IgA antibodies to desmogleins 1 and 3 in pemphigus vulgaris, pemphigus foliaceus and paraneoplastic pemphigus. Br J Dermatol. 2007 Apr;156(4):635-41. doi: 10.1111/j.1365-2133.2006.07717.x. Epub 2007 Jan 30. PMID: 17263817. https://pubmed.ncbi.nlm.nih.gov/17263817/
- Porro AM, Caetano Lde V, Maehara Lde S, Enokihara MM. Non-classical forms of pemphigus: pemphigus herpetiformis, IgA pemphigus, paraneoplastic pemphigus and IgG/IgA pemphigus. An Bras Dermatol. 2014 Jan-Feb;89(1):96-106. doi: 10.1590/abd1806-4841.20142459. PMID: 24626654; PMCID: PMC3938360. https://pubmed.ncbi.nlm.nih.gov/24626654/
- Hashimoto T, Teye K, Hashimoto K, et al. Clinical and Immunological Study of 30 Cases With Both IgG and IgA Anti-Keratinocyte Cell Surface Autoantibodies Toward the Definition of Intercellular IgG/IgA Dermatosis. Front Immunol. 2018 May 7;9:994. doi: 10.3389/fimmu.2018.00994. PMID: 29867971; PMCID: PMC5950707. https://pubmed.ncbi.nlm.nih.gov/29867971/
- Criscito MC, Cohen JM, Toosi S, et al. A retrospective study on the clinicopathologic features of IgG/IgA pemphigus. J Am Acad Dermatol. 2021 Jul;85(1):237-240. doi: 10.1016/j.jaad.2020.07.126. Epub 2020 Aug 13. PMID: 32798577.

https://pubmed.ncbi.nlm.nih.gov/32798577/

TESTING METHODS
Indirect Immunofluorescence (IIF)

IgA Epithelial Cell Surface (CS)/Intercellular Substance (ICS) Antibodies

Patient serum is progressively diluted in calcium-containing buffer beginning at 1:5 in three two-fold screening dilutions, layered on sections of intact normal human skin and monkey esophagus substrates, and reacted with fluorescein isothiocyanate (FITC)-conjugated antibody to IgA. When positive, the serum is further diluted in two-fold reductions to the limiting dilution of antibody detection or to a maximum dilution of 1:40,960. The limiting-dilution, end-point titer is reported for each substrate. This indirect immunofluorescence testing was developed, and its performance characteristics determined by the Immunodermatology Laboratory at the University of Utah. It has not been cleared or approved by the FDA (US Food and Drug Administration). FDA clearance or approval currently is not required for this testing performed in a CLIA-certified laboratory (Clinical Laboratory Improvement Amendments) and intended for clinical use. [Indirect immunofluorescence assays, one antibody on two substrates (IIF X 2) with two limiting dilution, end-point titers (antibody titer X 2)]

Electronically signed by [redacted] on 11/15/23 at 10:07 PM.

Resulting Laboratory

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