



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: [REDACTED]

ARUP Test Code: 0090650
Collection Date: 10/13/2023
Received in lab: 10/13/2023
Completion Date: 10/23/2023

Pemphigus Antibody Panel, IgG

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



Patient: [REDACTED]
ARUP Accession: 23-286-103711



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IMMUNODERMATOLOGY LABORATORY REPORT

Submitter

ARUP Sendouts

Pemphigus Antibody Panel, IgG (Final result)

TESTING REPORT follows "See Note"
See Note

CLINICAL INFORMATION

Mucosal erosions and scattered eroded skin lesions on upper body.
Presumptive diagnosis is pemphigus versus pemphigoid.

Specimen Details

- Serum; Collected: 10/13/2023; Received: 10/17/2023

DIAGNOSTIC INTERPRETATION

Consistent with pemphigus vulgaris

(See Results and Comments)

RESULTS

Indirect Immunofluorescence (IIF)

Cell Surface (CS)/Intercellular Substance (ICS) IgG Antibodies

IgG: Positive, titer 1:5120 (H), monkey esophagus
substrate

Positive, titer 1:2560 (H), intact human skin
substrate

Copy For:
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Printed: 10/23/2023 10:02 PM
Page: 1 of 4



Patient:
ARUP Accession: 23-286-103711

PCP: Unspecified

Reference Range:

Negative - Titer less than 1:10

Borderline - Titer 1:10

Positive (H) - Titer greater than 1:10

(H) = high/positive

Enzyme-Linked Immunosorbent Assay (ELISA)

Desmoglein (DSG) 1 and 3 IgG Antibodies

IgG desmoglein 1 antibody level: 3 U/mL

Reference Range:

Normal (negative) = Less than 14 U/mL

Borderline/Indeterminate = 14-20 U/mL

Increased (H) (positive) = Greater than 20 U/mL

IgG desmoglein 3 antibody level: 98 U/mL (H)

Reference Range:

Normal (negative) = Less than 9 U/mL

Borderline/Indeterminate = 9-20 U/mL

Increased (H) (positive) = Greater than 20 U/mL

(H) = high/positive

U = antibody level in ELISA units

COMMENTS

Specific

The indirect immunofluorescence results, demonstrating positive IgG cell surface (CS), also known as intercellular substance (ICS), antibodies, support the diagnosis of pemphigus. The ELISA results, demonstrating an increased IgG desmoglein 3 antibody level and a normal IgG desmoglein 1 antibody level, further support the diagnosis of pemphigus vulgaris. Serum IgG CS/ICS antibody titers by indirect immunofluorescence and IgG desmoglein antibody levels by ELISA correlate with disease activity in patients with IgG pemphigus variants.

IgG CS/ICS antibodies characteristically are positive by indirect immunofluorescence in IgG pemphigus variants, including pemphigus foliaceus and pemphigus vulgaris, and IgA CS/ICS antibodies characteristically are positive by indirect immunofluorescence in IgA pemphigus, although IgA CS/ICS antibodies may be observed in some pemphigus variants along with positive IgG CS/ICS antibodies. If indicated to further evaluate for IgA CS/ICS serum antibodies, add-on testing may be requested on this specimen by contacting ARUP Client Services at 1-800-242-2787, option 2, for:

Copy For:
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Printed: 10/23/2023 10:02 PM
Page: 2 of 4



Patient: [REDACTED]
ARUP Accession: 23-286-103711

PCP: Unspecified

- Pemphigus Antibodies, IgA by IIF (ARUP test number 0092106).]

Clinical correlation is needed, including treatment status, with consideration for monitoring serum antibody profiles by indirect immunofluorescence and antibody levels by ELISAs to aid in assessing disease expression and activity, including response to therapy.

General

More than 80 percent of patients with pemphigus have positive epithelial cell surface (CS) antibodies, also known as intercellular substance (ICS) antibodies, in their sera identified by indirect immunofluorescence. Serum antibody titers correlate with disease activity, and CS/ICS antibodies may be in low titer or negative in patients whose disease activity is minimal and/or under therapeutic control. CS/ICS antibodies are implicated in the pathophysiology of pemphigus. CS/ICS antibodies are typically not detected in normal individuals or in patients with other immunobullous diseases, 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. IgG CS/ICS antibodies characteristically are positive by indirect immunofluorescence in IgG pemphigus variants, including pemphigus foliaceus and pemphigus vulgaris. IgA CS/ICS antibodies are positive by indirect immunofluorescence in patients with IgA pemphigus and in some pemphigus variants along with positive IgG CS/ICS antibodies. Approximately 40 percent of patients with nonclassical IgG/IgA pemphigus have an underlying systemic disease when diagnosed, malignancy being the most common.

Pathogenic antibodies in serum from individuals with pemphigus bind to desmogleins, which are calcium-dependent adhesion molecules in epithelial desmosomes; such antibodies are detected by ELISA displaying principal desmoglein epitopes. Specific reactivity to the type of desmoglein may be helpful in determining pemphigus subtypes; IgG desmoglein 1 autoantibodies predominate in patients with pemphigus foliaceus, and IgG desmoglein 3 autoantibodies, with or without accompanying desmoglein 1 autoantibodies, predominate in patients with pemphigus vulgaris. Autoantibody expression to both desmogleins 1 and 3 is associated with both skin and mucosal lesions, often with clinical features of pemphigus foliaceus and pemphigus vulgaris. ELISA testing for IgG desmoglein 1 and IgG desmoglein 3 antibodies is highly sensitive, with greater than 90 percent of patients with IgG-variant pemphigus showing increased levels of one or both antibodies. IgG desmoglein antibody levels also correlate with disease activity in pemphigus foliaceus and pemphigus vulgaris; however, patients with cell surface/intercellular substance antibody-positive pemphigus by indirect immunofluorescence can have normal results on ELISA testing with antibodies to different desmoglein 1 and/or desmoglein 3 epitopes than in the ELISAs or to other desmosomal adhesion molecules.

Copy For:
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Printed: 10/23/2023 10:02 PM
Page: 3 of 4



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PCP: Unspecified

TESTING METHODS

Indirect Immunofluorescence (IIF)

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

Patient serum is progressively diluted in calcium-containing buffer beginning at 1:10 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 IgG. 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 IIF 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)]

Enzyme-Linked Immunosorbent Assays (ELISA)

IgG desmoglein 1 and IgG desmoglein 3 serum antibody levels determined by U.S. Food and Drug Administration (FDA)-approved ELISAs (Mesacup, MBL BION). [Two ELISAs]

Electronically signed by [REDACTED], on 10/23/23 at 10:02 PM.

Resulting Laboratory

| | |
|------------------------------------|--------------|
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| 417 S. Wakara Way, Suite 2151 | |
| Salt Lake City, UT 84108 | |
| Director: Kristin M. Leiferman, MD | |

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Printed: 10/23/2023 10:02 PM
Page: 4 of 4



Patient: [REDACTED]
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