ARUP LABORATORIES
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Jonathan R. Genzen, MD, PhD, Chief Medical Officer

Client: Example Client ABC123
123 Test Drive
Salt Lake City, UT 84108
UNITED STATES

Physician: Doctor, Example

Patient: Patient, Example

DOB	10/12/1995
Gender:	Female
Patient Identifiers:	01234567890ABCD, 012345
Visit Number (FIN):	01234567890ABCD
Collection Date:	00/00/0000 00:00

Pemphigus Antibody Panel, IgG

ARUP test code 0090650

EER Pemphigus Antibody Panel, IgG

See Note

Authorized individuals can access the ARUP Enhanced Report using the following link:

[redacted]

Pemphigus Antibody Panel, IgG

See Note

CLINICAL INFORMATION
Oral lesions and occasional skin erosions, possible pemphigus.

Specimen Details
[redacted] - Serum; Collected: 10/13/2023; Received: 10/17/2023

DIAGNOSTIC INTERPRETATION

Negative/normal IgG Pemphigus Antibody Panel

(See Results and Comments including further testing considerations)

RESULTS
Indirect Immunofluorescence (IIF)

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

IgG: Negative, monkey esophagus substrate

Negative, intact human skin substrate

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: 8 U/mL

Reference Range:
Normal (negative) = Less than 14 U/mL

H=High, L=Low, *=Abnormal, C=Critical

Borderline/Indeterminate = 14-20 U/mL
Increased (H) (positive) = Greater than 20 U/mL

IgG desmoglein 3 antibody level: 3 U/mL

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 negative IgG cell surface (CS), also known as intercellular substance (ICS), antibodies, by indirect immunofluorescence testing and normal IgG desmoglein 1 and IgG desmoglein 3 antibody levels by ELISAs are against, but do not rule out, the diagnosis of active pemphigus vulgaris, pemphigus foliaceus, or other IgG pemphigus variants. These findings do not rule out other immunobullous diseases.

If indicated to further evaluate the immunopathological profile for IgA CS/ICS antibodies as found in a rare variant of pemphigus, IgA pemphigus, and/or for basement membrane zone antibodies as found in pemphigoid, epidermolysis bullosa acquisita, or linear IgA disease, additional testing may be performed on this serum specimen by contacting ARUP Client Services at 1-800-242-2787, option 2, with add-on test request(s) for:
- Pemphigus Antibodies, IgA by IIF (ARUP test number 0092106); and/or
- Basement Membrane Zone Antibody Panel (ARUP test number 3001410).

Detection, levels, and patterns of diagnostic antibodies may fluctuate with disease manifestations. Clinical correlation is needed, including with direct immunofluorescence findings on a biopsy specimen and 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, particularly with persistent, progressive, or changing disease.

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. 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.

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 indirect immunofluorescence testing was developed and its performance characteristics were 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)]

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 10:05 PM.
Performed At: IMMUNODERMATOLOGY LABORATORY
417 S. WAKARA WAY, SUITE 2151
SALT LAKE CITY, UT 84108
Medical Director: KRISTIN M. LEIFERMAN, MD
CLIA Number: 46D0681916

H=High, L=Low, *=Abnormal, C=Critical

VERIFIED/REPORTED DATES

Procedure	Accession	Collected	Received	Verified/Reported
EER Pemphigus Antibody Panel, IgG	23-286-107171	00/00/0000 00:00	00/00/0000 00:00	00/00/0000 00:00
Pemphigus Antibody Panel, IgG	23-286-107171	00/00/0000 00:00	00/00/0000 00:00	00/00/0000 00:00

END OF CHART

H=High, L=Low, *=Abnormal, C=Critical