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	10/12/1980
Gender:	Female
Patient Identifiers:	01234567890ABCD, 012345
Visit Number (FIN):	01234567890ABCD
Collection Date:	00/00/0000 00:00

Cell Surface (Epithelial) Antibodies, IgG by IIF

ARUP test code 0090266

EER Epithelial Cell Surface Antibody IgG

See Note

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

[redacted]

Epithelial Cell Surface Ab IgG

See Note

CLINICAL INFORMATION
Mucosal erosions and few scattered eroded skin lesions on upper body. Presumptive diagnosis is pemphigus versus mucous membrane pemphigoid.

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

DIAGNOSTIC INTERPRETATION

Consistent with IgG-variant pemphigus, including pemphigus foliaceus and pemphigus vulgaris

(See Results and Comments including further testing recommendations)

RESULTS
Indirect Immunofluorescence (IIF)

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

IgG: Positive, titer 1:2560 (H), monkey esophagus substrate
Positive, titer 1:640 (H), 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

COMMENTS
Specific

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

These indirect immunofluorescence results, demonstrating positive IgG cell surface (CS), also known as intercellular substance (ICS), antibodies, reacting with both monkey esophagus substrate and intact human skin substrate, support the diagnosis of pemphigus vulgaris or pemphigus foliaceus or another IgG-variant pemphigus. Pemphigus foliaceus and pemphigus vulgaris can be distinguished by the predominance of IgG desmoglein 1 or IgG desmoglein 3 antibodies, respectively, determined by enzyme-linked immunosorbent assay (ELISA). Another consideration is that, although IgA CS/ICS antibodies characterize IgA pemphigus, they also may be observed in some pemphigus variants along with positive IgG CS/ICS antibodies.

Further ELISA testing for IgG desmoglein 1 and IgG desmoglein 3 antibody levels is recommended and can be accomplished on this specimen by contacting ARUP Client Services, 1-800-242-2787, option 2, with add-on test request for:
- Desmoglein 1 and Desmoglein 3 (Pemphigus) Antibodies, IgG by ELISA (ARUP test number 0090649),

with or without additional indirect immunofluorescence testing to determine if IgA CS/ICS antibodies also are expressed with add-on test request for:
- Pemphigus Antibodies, IgA by IIF (ARUP test number 0092106).

IgG CS/ICS antibody titers and IgG desmoglein antibody levels correlate with disease activity in IgG-variant pemphigus, including pemphigus foliaceus and pemphigus vulgaris. Of note, cell surface reactivity by indirect immunofluorescence may be observed transiently and/or nonspecifically in normal individuals, and in patients with infections, drug reactions, and other mucocutaneous disorders. Clinical correlation is needed, including treatment status and with direct immunofluorescence findings on a biopsy specimen. Monitoring antibody levels by ELISAs as well as antibody profiles by indirect immunofluorescence can 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 typically are 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. Specific reactivity to the type of desmoglein, determined by enzyme-linked immunosorbent assay (ELISA) may be helpful in determining pemphigus subtypes; IgG desmoglein 1 antibody levels are increased in patients with pemphigus foliaceus, and IgG desmoglein 3 antibody levels with or without increased IgG desmoglein 1 antibody levels, are predominantly increased in patients with 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.

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 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, one antibody on two substrates (IIF X 2) with two limiting-dilution, end-point titers (antibody titer X 2)]

Electronically signed by [redacted], on 10/23/23 at 10:07 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

VERIFIED/REPORTED DATES

Procedure	Accession	Collected	Received	Verified/Reported
EER Epithelial Cell Surface Antibody IgG	23-286-103691	00/00/0000 00:00	00/00/0000 00:00	00/00/0000 00:00
Epithelial Cell Surface Ab IgG	23-286-103691	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