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

Physician: Doctor, Example

Patient: Patient, Example

DOB	12/31/1993
Gender:	Female
Patient Identifiers:	01234567890ABCD, 012345
Visit Number (FIN):	01234567890ABCD
Collection Date:	00/00/0000 00:00

Basement Membrane Zone and Cell Surface (Epithelial) Antibodies, IgG and IgA by IIF

ARUP test code 0090299

EER Epithelial Skin Antibodies

See Note

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

[redacted]

Epithelial Skin Antibodies

See Note

CLINICAL INFORMATION
Erythematous and urticarial plaques.

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

DIAGNOSTIC INTERPRETATION

Negative IgG, IgG4, and IgA basement membrane zone and cell surface/intercellular substance epithelial antibodies by indirect immunofluorescence

(See Results and Comments including further testing recommendations)

RESULTS
Indirect Immunofluorescence (IIF)

Basement Membrane Zone (BMZ) IgG, IgG4, and IgA Antibodies

IgG: Negative, monkey esophagus substrate
Negative, human split skin substrate

IgG4: Negative, monkey esophagus substrate
Negative, human split skin substrate

IgA: Negative, monkey esophagus substrate
Negative, human split skin substrate

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

Localization Pattern on Human BMZ Split Skin:
Epidermal (roof) or combined epidermal-dermal (roof and floor) IgG and/or IgG4 BMZ antibodies = pemphigoid (including pemphigoid gestationis,

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

bullous pemphigoid, some types of mucous membrane pemphigoid)

Dermal (floor) IgG and/or IgG4 BMZ antibodies = epidermolysis bullosa acquisita or bullous lupus erythematosus or anti-laminin-332 pemphigoid or anti-p200 (laminin gamma-1) pemphigoid or another rare pemphigoid subtype

Epidermal (roof), combined epidermal-dermal (roof and floor), or dermal (floor) IgA BMZ antibodies = linear IgA disease (including linear IgA bullous dermatosis and chronic bullous disease of childhood)

IgA and IgG basement membrane zone antibodies may be co-expressed in basement membrane zone antibody-associated diseases

(H) = high/positive

Cell Surface (CS)/Intercellular Substance (ICS) IgG, IgG4, and IgA
Antibodies

IgG: Negative, monkey esophagus substrate
Negative, intact human skin substrate

IgG4: Negative, monkey esophagus substrate
Negative, intact human skin substrate

IgA: 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

COMMENTS

Specific

The negative IgG, including IgG4, and IgA basement membrane zone antibodies by indirect immunofluorescence testing do not provide support for, but do not rule out, the diagnoses of pemphigoid, epidermolysis bullosa acquisita, and linear IgA disease. In certain patients with pemphigoid, IgG BP180 and/or IgG BP230 antibody levels by ELISAs may be more sensitive diagnostic markers than indirect immunofluorescence, and, in certain patients with epidermolysis bullosa acquisita, the IgG type VII collagen antibody level by ELISA may be a more sensitive diagnostic marker than indirect immunofluorescence. Therefore, if clinically indicated to further evaluate for pemphigoid and/or epidermolysis bullosa acquisita, ELISA testing may be accomplished on this specimen by add-on test request for these ELISAs; see below for ordering and contact information.

The negative IgG, including IgG4, and IgA cell surface (CS), also known as intercellular substance (ICS), antibodies by indirect immunofluorescence testing do not provide support for, but do not rule out, the diagnoses of pemphigus vulgaris, pemphigus foliaceus, other IgG pemphigus variants, and IgA pemphigus. In certain patients with pemphigus, IgG desmoglein 1 and/or IgG desmoglein 3 antibody levels by ELISAs may be more sensitive diagnostic markers than indirect immunofluorescence. Therefore, if clinically indicated to further evaluate for pemphigus foliaceus and pemphigus vulgaris, ELISA testing may be

accomplished on this specimen by add-on test request for these ELISAs; see below for ordering and contact information.

Additional testing on this serum specimen may be requested by contacting ARUP Client Services at 1-800-242-2787, option 2, with add-on test request(s) for:
- Bullous Pemphigoid (BP180 and BP230) Antibodies, IgG by ELISAs (ARUP test number 0092566),
- Collagen Type VII Antibody, IgG by ELISA (ARUP test number 2010905), and/or
- Desmoglein 1 and Desmoglein 3 (Pemphigus) Antibodies, IgG by ELISA (ARUP test number 0090649).

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 levels by ELISAs along with antibody profiles by indirect immunofluorescence to aid in assessing disease expression and activity.

General

Approximately 80 percent of patients with bullous pemphigoid, epidermolysis bullosa acquisita, and linear IgA bullous dermatosis have positive antibodies to basement membrane zone components in their sera detected by indirect immunofluorescence. Approximately 50 percent of patients with mucous membrane/cicatricial pemphigoid demonstrate antibodies to basement membrane zone components detected by indirect immunofluorescence. The immunoglobulin class of basement membrane zone antibodies and pattern of antibody localization on split skin substrate distinguish the diseases. IgG4 subclass reactivity by indirect immunofluorescence may be more sensitive than IgG in some patients with pemphigoid and epidermolysis bullosa acquisita. Positive serum IgA epithelial basement membrane zone antibodies are highly specific diagnostic markers for linear IgA disease. IgA basement membrane zone antibodies by indirect immunofluorescence may be found in variant presentations of mucous membrane pemphigoid and epidermolysis bullosa acquisita. Moreover, IgA basement membrane zone antibodies may be co-expressed with IgG basement membrane zone antibodies in some patients with pemphigoid including mucous membrane/cicatricial pemphigoid and in linear IgA/IgG bullous dermatosis.

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. IgG4 subclass reactivity by indirect immunofluorescence may be more sensitive than IgG in some patients with pemphigus. 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. 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.

TESTING METHODS
Indirect Immunofluorescence (IIF)

IgG, IgG4, and IgA Epithelial Basement Membrane Zone (BMZ) and

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

Patient serum is progressively diluted beginning at 1:5 in four two-fold screening dilutions, layered on sections of human skin split at the basement membrane zone, intact human skin, and monkey esophagus substrates, and reacted with fluorescein isothiocyanate (FITC)-conjugated antibodies to IgG and 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, and the pattern of staining on split skin substrate also is reported. FITC-conjugated anti-IgG4 is tested to increase test sensitivity (maximum serum dilution of 1:40). 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, three antibodies on three substrates (IIF X 9)]

Electronically signed by [redacted] on 11/15/23 at 9:59 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 Skin Antibodies	23-307-104783	00/00/0000 00:00	00/00/0000 00:00	00/00/0000 00:00
Epithelial Skin Antibodies	23-307-104783	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