Client: Example Client ABC123
123 Test Drive

UNITED STATES

Physician: Doctor, Example

Patient: Patient, Example

DOB

Sex:

Patient Identifiers: 01234567890ABCD, 012345

Visit Number (FIN): 01234567890ABCD

Collection Date: 01/01/2017 12:34

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

ARUP test code 0090299

Epithelial Skin Antibodies

See Note

CLINICAL INFORMATION
Erythematous and urticarial plaques.

Specimen Details
S22-IP0000524 - Serum; Collected: ; Received:

DIAGNOSTIC INTERPRETATION

Negative IgG and IgA epithelial basement membrane zone and cell surface antibodies by indirect immunofluorescence

(See Results and Comments including further testing recommendations)

RESULTS
Indirect Immunofluorescence (IIF)

Basement Membrane Zone (BMZ) IgG and IgA Antibodies

IgG: 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, bullous pemphigoid, 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)

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

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

IgG: 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 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. If clinically indicated to further evaluate for pemphigoid and/or epidermolysis bullosa acquisita, additional testing may be accomplished on this specimen by add-on test request for IgG BP180, IgG BP230, IgG collagen VII antibody ELISAs; see below for ordering and contact information.

The negative IgG and IgA cell surface antibodies by indirect immunofluorescence testing do not provide support for, but do not rule out, the diagnoses of pemphigus vulgaris, pemphigus foliaceus, other types of IgG pemphigus, 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. If clinically indicated to further evaluate for pemphigus foliaceus and pemphigus vulgaris, additional testing may be accomplished on this specimen by add-on test request for IgG desmoglein 1 and/or IgG desmoglein 3 antibody ELISAs; see below for ordering and contact information.

Add-on testing may be requested on this serum specimen by contacting ARUP Client Services at 1-800-242-2787, option 2, 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. 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 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. 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, two antibodies on three substrates (IIF X 6)]

Electronically signed by , MD, on
at .
Performed At:

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

Medical Director: , MD
CLIA Number:

VERIFIED/REPORTED DATES

Procedure	Accession	Collected	Received	Verified/Reported
Epithelial Skin Antibodies	22-173-114819			

END OF CHART

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