

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

Physician: Doctor, Example

Patient: Patient, Example

DOB: 01/01/1990
Gender: Female
Patient Identifiers: 01234567890ABCD, 012345
Visit Number (FIN): 01234567890ABCD
Collection Date: 01/01/2017 12:34

Pemphigoid Antibody Panel - Epithelial Basement Membrane Zone Antibodies, IgG and IgA, BP180 and BP230 Antibodies, IgG

ARUP test code 0092001

Pemphigoid Antibody Panel

See Note
IMMUNODERMATOLOGY REPORT

Specimen(s):
1. Serum specimen

Clinical/Diagnostic Information:
No clinical information provided.

DIAGNOSTIC INTERPRETATION

Negative/normal Pemphigoid Panel antibodies

(See Results and Comments including additional test consideration)

RESULTS

Indirect Immunofluorescence

Basement Membrane Zone (BMZ) IgG 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:

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

Pattern on Human BMZ Split Skin:

IgG epidermal or epidermal-dermal combined BMZ
antibody pattern = pemphigoid

IgG dermal BMZ antibody pattern = epidermolysis
bullosa acquisita

IgA epidermal, epidermal-dermal combined, or,
dermal BMZ antibody pattern = linear IgA bullous
dermatosis

(H = high/positive)

H - high L - low * - abnormal C - critical

Enzyme Linked Immunosorbent Assay (ELISA)

Bullous Pemphigoid (BP) 180 and 230 IgG Antibodies

IgG BP 180 antibodies: 0 units

Reference Range:

Positive (H) = Greater than or equal to 9 units

Negative = Less than 9 units

IgG BP 230 antibodies: 2 units

Reference Range:

Positive (H) = Greater than or equal to 9 units

Negative = Less than 9 units

(H = high/increased; units = units/mL serum)

COMMENTS

Specific

These negative/normal results for basement membrane zone antibodies, including IgG4, by indirect immunofluorescence and ELISA testing are against, but do not rule out, the diagnoses of bullous pemphigoid, epidermolysis bullosa acquisita, and linear IgA bullous dermatosis. The results do not rule out the diagnosis of mucous membrane/cicatricial pemphigoid because a large proportion of patients with this pemphigoid subtype do not have detectable circulating basement membrane zone antibodies.

IgG Collagen VII antibodies by ELISA may be a more sensitive diagnostic marker than indirect immunofluorescence in some patients with epidermolysis bullosa acquisita. Therefore, if epidermolysis bullosa acquisita is a diagnostic consideration, recommend additional testing for IgG Collagen VII antibodies by ELISA (ARUP test number 2010905), which may be performed on this specimen by add-on test request through ARUP Client Services at 1-800-242-2787 option 2.

Recommend monitoring antibody profiles over time for persistent or progressive disease activity. If it would be helpful to discuss this case with these results and/or further testing, contact ARUP Client Services at 1-800-242-2787 option 2 and ask a representative to connect you with the Immunodermatology Laboratory at the University of Utah.

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. Approximately 20 percent of patients with mucous membrane pemphigoid, including cicatricial/mucous membrane pemphigoid, demonstrate antibodies to basement membrane zone components. The pattern of staining on split skin specifies disease. IgG4 subclass reactivity may be more sensitive than IgG in some immunobullous disease cases.

Major molecular structures in the basement membrane zone to which IgG pemphigoid antibodies bind have been identified and termed "BP 180" for a 180 kDa bullous pemphigoid antigen and "BP 230" for a 230 kDa bullous pemphigoid antigen. BP 180 is a transmembrane component of the basement membrane zone with collagen-like domains and is a principal antigenic target in mucous membrane pemphigoid. BP 230 is located in the hemidesmosomal plaque of basal cells in the epidermis. Serum levels of IgG BP 180 and IgG BP 230 antibodies are in the negative range in normal individuals, and serum levels of IgG BP 180 may correlate with disease activity in pemphigoid, diminishing with treatment response. Patients with pemphigoid may show reactivity to multiple basement membrane zone components in addition to or other than the BP 180 and BP 230 epitopes expressed in these ELISAs.

TESTING METHODS

H – high L – low * – abnormal C – critical

Indirect Immunofluorescence

 Basement Membrane Zone (BMZ) IgG and IgA Antibodies

The patients serum is progressively diluted beginning at 1:5 in three two-fold screening dilutions, layered on sections of monkey esophagus substrate and human basement membrane zone split skin substrate, and stained with fluorescein-conjugated anti-IgA and anti-IgG using Analyte Specific Reagents (ASRs). 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. If indicated, fluorescein-conjugated anti-IgG4 also is tested to increase test sensitivity (maximum serum dilution of 1:20). These tests were developed and their performance characteristics determined by the Immunodermatology Laboratory at the University of Utah. They have not been cleared or approved by the U.S. Food and Drug Administration. ASRs are used in many laboratory tests necessary for standard medical care and generally do not require FDA approval. These tests should not be regarded as investigational or for research only. [Immunofluorescence studies, three antibodies on two substrates]

Enzyme Linked Immunosorbent Assay (ELISA)

 IgG BP 180 and IgG BP 230 serum antibody levels determined by U.S. Food and Drug Administration-approved ELISAs (Mesacup, MBL International). [Two ELISAs]

Kristin M Leiferman, MD
 Immunodermatologist
 Electronically signed 1/12/2017 9:29:20PM
 Performed at: ARUP - University Hospital Laboratory 50 N. Medical Drive Salt Lake City UT 84132

EER Pemphigoid Antibody Panel

See Note

Access ARUP Enhanced Report using either link below:

-Direct access: [REDACTED]

-Enter Username, Password: [REDACTED]

Username: [REDACTED]
 Password: [REDACTED]
 Performed at: ARUP - University Hospital Laboratory 50 N. Medical Drive Salt Lake City UT 84132

VERIFIED/REPORTED DATES

Procedure	Accession	Collected	Received	Verified/Reported
Pemphigoid Antibody Panel	17-010-403295	1/10/2017 11:36:00 AM	1/11/2017 4:00:25 AM	1/13/2017 11:59:40 AM
EER Pemphigoid Antibody Panel	17-010-403295	1/10/2017 11:36:00 AM	1/11/2017 4:00:25 AM	1/13/2017 11:59:40 AM

END OF CHART

H - high L - low * - abnormal C - critical