

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

Consistent with pemphigoid with typical, predominant IgG basement membrane zone antibodies and also with IgA basement membrane zone antibodies

(See Results, Comments, separate concurrent Epithelial Skin Antibody report with positive findings and additional comments, and Concurrent Test Results Chart)

RESULTS

Indirect Immunofluorescence

Basement Membrane Zone (BMZ) IgG and IgA Antibodies

IgG: Positive, titer 1:5120 (H), monkey esophagus substrate
Positive, epidermal-dermal combined pattern, epidermal
Predominant, titer 1:1280 (H), human split
skin substrate

IgA: Positive, titer 1:80 (H), monkey esophagus substrate
Positive, epidermal pattern, titer 1:160 (H), 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

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

dermatosis

(H = high/positive)

Enzyme Linked Immunosorbent Assay (ELISA)

Bullous Pemphigoid (BP) 180 and 230 IgG Antibodies

IgG BP 180 antibodies: 163* units (H)

Reference Range:

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

IgG BP 230 antibodies: 18 units (H)

Reference Range:

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

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

* This is an extrapolated value, greater than the highest calibrator in the assay. This level is markedly increased; however, this antibody level is not on the linear portion of the calibrator curve and may not be comparable to other levels above the calibrator range or to levels within calibrator range, 0-100 units/mL. If it would be clinically helpful in monitoring disease activity, dilution testing of this specimen may be requested to obtain comparable levels of antibodies in the high ranges. Contact ARUP Client Services at 801-583-2787 (or toll free at 1-800-242-2787) option 2 and ask a representative to connect you with the Immunodermatology Laboratory at the University of Utah.

COMMENTS

Specific

In addition to positive IgG basement membrane zone antibodies, characteristic of pemphigoid, positive IgA basement membrane zone antibodies, characteristic of linear IgA bullous dermatosis, are found in this patient's serum. The predominance of the IgG basement membrane zone antibodies favors the diagnosis of pemphigoid over linear IgA bullous dermatosis; however, the presence of two antibody classes reactive with basement membrane zone may have implications for disease severity and treatment considerations.

IgG BP 180 antibody levels correlate with disease activity in some cases of pemphigoid; therefore, testing antibody levels over time may be useful in monitoring disease activity and response to therapy. Monitoring antibody profiles with respect to IgG and IgA basement membrane zone reactivity also may be useful in monitoring disease expression.

See separate, concurrent Epithelial Skin Antibody report for positive findings with additional comments, and see chart for concurrent test results.

CONCURRENT TEST RESULTS CHART

Basement Membrane Zone Antibodies

Serum Number	Date of Specimen	IgG BMZ Titers	IgA BMZ Titers	BP 180	BP 230	Coll VII
██████████	01/05/17	ME: 1:5120 SS: Combined, Epi Predominant, 1:1280	ME: 1:80 SS: Epi, 1:160	163*	18	NA
██████████ (See Note 1)	01/05/17	ME: 1:5120 SS: Combined, Epi Predominant, 1:2560	ME: 1:80 SS: Epi, 1:160	NA	NA	NA

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

Note 1: Negative IgG and IgA cell surface antibodies by indirect immunofluorescence in Epithelial Skin Antibody testing.

Chart Key:

IgG BMZ = IgG basement membrane zone antibodies by indirect immunofluorescence
 IgA BMZ = IgA basement membrane zone antibodies by indirect immunofluorescence

ME = Antibody absence (negative) or presence with endpoint titer on monkey esophagus substrate

SS = Antibody absence (negative) or presence with endpoint titer and pattern on split skin substrate

Epi = epidermal staining pattern on split skin substrate

Dermal = dermal staining pattern on split skin substrate

Combined = combined epidermal-dermal staining pattern

BP 180 = IgG BP 180 antibody level (Units/mL) by ELISA

BP 230 = IgG BP 230 antibody level (Units/mL) by ELISA

Coll VII = IgG Collagen VII antibody level (Units/mL) by ELISA

NA = Not Assayed

COMMENTS

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 have positive antibodies to basement membrane zone components in their sera. The pattern of staining on split skin specifies disease.

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. 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 IgG BP 180 antibody levels correlate with disease activity in some cases of pemphigoid. 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

Indirect Immunofluorescence

Basement Membrane Zone (BMZ) IgG and IgA Antibodies

The patient's 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. 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, two antibodies on two substrates]

Enzyme Linked Immunosorbent Assay (ELISA)

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

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 11:27:03AM
 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	17-006-400587	1/5/2017 11:47:00 AM	1/9/2017 2:26:33 PM	1/13/2017 11:43:20 AM
EER Pemphigoid Antibody Panel	17-006-400587	1/5/2017 11:47:00 AM	1/9/2017 2:26:33 PM	1/13/2017 11:43:20 AM

END OF CHART

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