Pemphigoid Antibody Panel - Epithelial Basement Membrane Zone Antibodies, IgG and IgA, BP180 and BP230 Antibodies, IgG
ARUP test code 0092001

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 bullosa

H - high  L - low  * - abnormal  C - critical
**Bullous Pemphigoid (BP) 180 and 230 IgG Antibodies**

<table>
<thead>
<tr>
<th>Serum Number</th>
<th>Date of Specimen</th>
<th>IgG BMZ Titers</th>
<th>IgA BMZ Titers</th>
<th>BP</th>
<th>BP</th>
<th>Coll VII</th>
</tr>
</thead>
<tbody>
<tr>
<td>01/05/17</td>
<td>ME: 1:5120</td>
<td>ME: 1:80</td>
<td>163*</td>
<td>18</td>
<td>NA</td>
<td></td>
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<tr>
<td></td>
<td>SS: Combined,</td>
<td>SS: Epi,</td>
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<tr>
<td></td>
<td>Epi Predominant,</td>
<td>1:1280</td>
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</table>

**Note 1:** 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.
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
- **BP180** = IgG BP180 antibody level (Units/mL) by ELISA
- **BP230** = IgG BP230 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 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. 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:
- Enter Username, Password:
  Username: [Redacted]
  Password: [Redacted]
Performed at: ARUP - University Hospital Laboratory 50 N. Medical Drive Salt Lake City UT 84132

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Accession</th>
<th>Collected</th>
<th>Received</th>
<th>Verified/Reported</th>
</tr>
</thead>
</table>

END OF CHART

H—high L—low *—abnormal C—critical