



# Basement Membrane Zone (Epithelial) Antibodies, IgG by IIF

## LABORATORIES

Patient: [REDACTED]  
DOB: [REDACTED] Age: [REDACTED]  
Patient Identifiers: [REDACTED]  
Visit Number (FIN): [REDACTED]

Sex: [REDACTED]

Client: ARUP Example Report Only  
500 Chipeta Way  
Salt Lake City, UT 84108  
Physician: [REDACTED]

ARUP Test Code: 0092056  
Collection Date: 11/17/2023  
Received in lab: 11/17/2023  
Completion Date: 11/17/2023

### Immunodermatology Serum Test Report Navigation Guide

The Immunodermatology TESTING REPORT from the University of Utah follows "See Note" and is arranged as outlined below on the following pages:

#### CLINICAL INFORMATION

This content is provided by the ordering clinician and includes the reason for testing.

#### Specimen Details

This includes specimen identification with collected and received dates.

#### DIAGNOSTIC INTERPRETATION

This is a synopsis of key findings from the testing and their diagnostic relevance.

#### RESULTS

This section reports the discrete finding and value of each test component, along with the reference range.

#### COMMENTS

Specific

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These comments provide an explanation of the test results as they relate to clinical considerations, and include reference to any concurrent and/or previous testing.

General

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These comments summarize fundamental information about the test(s) and the component(s) assessed to aid in interpretation of their clinical applicability.

#### TESTING METHODS

The section lists the procedures performed, the test source(s), and the applicable laboratory developed test disclaimer(s).

#### TEST RESULTS SUMMARY CHART

A chart tabulating results of tests ordered for the patient by the same client is included if previous and/or concurrent testing has been performed.

#### ELISA RESULTS GRAPH

A graph of ELISA results also is included if previous and/or concurrent testing has been performed; the graph may be found on a subsequent page.

For testing algorithm and additional information, refer to: [arupconsult.com/content/immunobullous-skin-diseases-screening](http://arupconsult.com/content/immunobullous-skin-diseases-screening)



Patient: [REDACTED]  
ARUP Accession: 23-321-114328



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## IMMUNODERMATOLOGY LABORATORY REPORT



Submitter

ARUP Sendouts

Basement Membrane Zone (Epithelial) Antibodies, IgG by IIF (Final result)

TESTING REPORT follows "See Note"  
See Note

### CLINICAL INFORMATION

Skin fragility, tense blisters, vesicles, erosions, and milia.  
Presumptive diagnosis is epidermolysis bullosa acquisita versus porphyria cutaneous tarda.

### Specimen Details

[Redacted] - ; Collected: 11/17/2023; Received: 11/17/2023

### DIAGNOSTIC INTERPRETATION

Positive IgG, including IgG4, basement membrane zone antibodies demonstrating dermal localization (floor) with split skin substrate by indirect immunofluorescence and concurrent ELISA testing demonstrating an increased IgG type VII collagen antibody level, consistent with subepidermal immunobullous disease, including epidermolysis bullosa acquisita or bullous lupus erythematosus

(See Results, Comments, separate concurrent Collagen Type VII Antibody, IgG by ELISA testing report with additional findings and comments, and Basement Membrane Zone Antibody Test Results Summary Chart with concurrent findings)

### RESULTS

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Patient: [Redacted]  
ARUP Accession: 23-321-114328

PCP: Unspecified

Indirect Immunofluorescence (IIF)

Basement Membrane Zone (BMZ) IgG and IgG4 Antibodies

IgG: Positive, titer 1:5120 (H), monkey esophagus substrate  
Positive, dermal pattern (floor), titer 1:2560 (H), human split skin substrate

IgG4: Positive, titer greater than 1:20 (H), monkey esophagus substrate  
Positive, dermal pattern (floor), titer greater than 1:20 (H), 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, 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

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

(H) = high/positive

COMMENTS

Specific

Based on the dermal localization (floor) of IgG, including IgG4, basement membrane zone antibodies with human split skin substrate (also known as salt split skin), these indirect immunofluorescence findings support the diagnosis of epidermolysis bullosa acquisita. In addition, the IgG type VII collagen antibody level is increased by ELISA in concurrent testing, further supporting the diagnosis of epidermolysis bullosa acquisita (separate report with additional comments). See chart (below) for summary of concurrent basement membrane zone antibody test

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results.

A subset of patients with bullous systemic lupus erythematosus has the same indirect immunofluorescence reactivity as epidermolysis bullosa acquisita with dermal (floor) IgG basement membrane zone antibody localization on split skin substrate and also may exhibit increased levels of IgG type VII collagen antibodies. Two subsets of pemphigoid, namely, anti-laminin-332 and anti-p200 (laminin gamma-1) pemphigoid, demonstrate IgG basement membrane zone antibody reactivity with the dermal side of the split skin substrate, although these two pemphigoid subsets do not characteristically demonstrate increased levels of IgG type VII collagen antibodies, as observed in this patient. Furthermore, IgG and IgA basement membrane zone antibodies may be co-expressed which may have implications for disease severity and treatment considerations. If indicated to further define the immunopathological profile with respect to basement membrane zone antibodies, additional testing may be performed on this serum specimen by contacting ARUP Client Services at 1-800-242-2787, option 2, with add-on test request for:

- Basement Membrane Zone (Epithelial) Antibodies, IgA by indirect immunofluorescence (ARUP test number 0092057).

Patients with inflammatory bowel disease, including Crohn disease and ulcerative colitis, with and without mucocutaneous manifestations of epidermolysis bullosa acquisita or associated with lupus erythematosus may demonstrate increased levels of IgG type VII collagen antibodies. Therefore, although the overall immunopathological profile is consistent with epidermolysis bullosa acquisita, the findings may be observed in bullous lupus erythematosus and do not rule out the diagnoses of anti-laminin-332 pemphigoid or anti-p200 (laminin gamma-1) pemphigoid with increased IgG type VII collagen antibodies associated with another condition.

Disorders with dermal IgG basement membrane zone antibodies on split skin substrate by indirect immunofluorescence cannot be further distinguished with currently available diagnostic laboratory techniques, except laminin-332 IgG antibodies in select laboratories. It is important to note that up to one third of patients with anti-laminin-332 pemphigoid have associated malignancy. Therefore, clinical correlation is needed with clinical evaluation as indicated. Monitoring serum antibody profiles by indirect immunofluorescence and antibody levels by ELISAs may aid in assessing disease expression and activity, including response to therapy.

If it would be helpful to discuss the patient case with this report, contact ARUP Client Services at 1-800-242-2787, option 2, and ask to speak with the Immunodermatology Laboratory at the University of Utah regarding patient results.

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General

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Approximately 80 percent of patients with bullous pemphigoid and epidermolysis bullosa acquisita have positive IgG antibodies to basement membrane zone components in their sera; the pattern of antibody localization on split skin substrate, also known as salt split skin, distinguishes the diseases. Approximately 50 percent of patients with mucous membrane/cicatrical pemphigoid demonstrate antibodies to basement membrane zone components. IgG4 subclass reactivity by indirect immunofluorescence may be more sensitive than IgG in some patients with immunobullous disease. IgA basement membrane zone antibodies may be co-expressed with IgG basement membrane zone antibodies in some patients with pemphigoid including mucous membrane/cicatrical pemphigoid, epidermolysis bullosa acquisita, lupus erythematosus, and linear IgA/IgG bullous dermatosis.

Major molecular structures in the basement membrane zone to which IgG pemphigoid antibodies bind have been identified and termed "BP180" for a 180 kDa bullous pemphigoid antigen and "BP230" for a 230 kDa bullous pemphigoid antigen. Type VII collagen is a component of anchoring fibrils within epithelial basement membrane zone (skin and mucous membranes) and is an antigenic target of IgG autoantibodies in patients with epidermolysis bullosa acquisita and in a subset of patients with bullous lupus erythematosus. Antibodies to these molecular structures are determined by enzyme-linked immunosorbent assay (ELISA). ELISA may be more sensitive testing than indirect immunofluorescence but may be more restricted than indirect immunofluorescence in assessing antibody reactivity to certain epitopes of the target components rather than the basement membrane zone in tissue.

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TESTING METHODS

Indirect Immunofluorescence (IIF)

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IgG and IgG4 Epithelial Basement Membrane Zone (BMZ) Antibodies

Patient serum is progressively diluted beginning at 1:5 in three two-fold screening dilutions, layered on sections of human skin split at the basement membrane zone and monkey esophagus substrates, and reacted with fluorescein isothiocyanate (FITC)-conjugated antibodies to IgG. 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 1:20). 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 two substrates (IIF X 4) with two limiting dilution, end-point titers

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(antibody titer X 2)]

TEST RESULTS SUMMARY CHART

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Basement Membrane Zone Antibodies

Serum Number	Date of Specimen	IgG and IgG4 BMZ Titers	IgA BMZ Titers	BP		Col VII
				180	230	
23-8601	11/03/23	IgG ME: 1:5120 IgG SS: Derm, 1:2560 IgG4 ME:>1:20 IgG4 SS: Derm, >1:20	ME: NA SS: NA	NA	NA	NA
23-8602	11/03/23	IgG ME: NA IgG SS: NA IgG4 ME: NA IgG4 SS: NA	ME: NA SS: NA	NA	NA	88

ELISA Reference Ranges:

IgG BP180 and IgG BP230 Antibody Levels  
Normal (negative) = Less than 9 U/mL  
Increased (H) (positive) = 9 U/mL and greater

IgG Type VII Collagen Antibody Level  
Normal (negative) = Less than 7 U/mL  
Slightly increased (H) (positive) = 7-8 U/mL  
Increased (H) (positive) = 9 U/mL and greater

Chart Key:

IgG BMZ = IgG basement membrane zone (BMZ) antibodies by indirect immunofluorescence  
IgG4 BMZ = IgG4 basement membrane zone (BMZ) antibodies by indirect immunofluorescence  
IgA BMZ = IgA basement membrane zone (BMZ) antibodies by indirect immunofluorescence

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

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

Epi = epidermal localization (roof) on split skin substrate (IgG - pemphigoid including bullous pemphigoid, some mucous membrane pemphigoid, and

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PCP: Unspecified

other pemphigoid variants; IgA - linear IgA disease including linear IgA bullous dermatosis and chronic bullous disease of childhood)  
Derm = dermal localization (floor) on split skin substrate (IgG - epidermolysis bullosa acquisita, bullous lupus erythematosus, anti-laminin-332 pemphigoid, anti-p200 (laminin gamma-1) pemphigoid, other rare pemphigoid subtypes; IgA - linear IgA disease including linear IgA epidermolysis bullosa acquisita)  
Comb = combined epidermal-dermal localization (roof and floor) on split skin substrate (IgG - pemphigoid and pemphigoid variants; IgA - linear IgA disease)

BP180 = IgG BP180 antibody level (U/mL) by ELISA  
BP230 = IgG BP230 antibody level (U/mL) by ELISA  
Col VII = IgG Collagen VII antibody level (U/mL) by ELISA

NA = Not Assayed

Electronically signed by [REDACTED] on 11/17/23 at 4:17 PM.

**Resulting Laboratory**

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