



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: 0092001
Collection Date: 03/19/2024
Received in lab: 03/19/2024
Completion Date: 03/22/2024

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

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

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



Patient: [REDACTED]
ARUP Accession: 24-079-103960



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IMMUNODERMATOLOGY LABORATORY REPORT

Submitter

ARUP Sendouts

Pemphigoid Antibody Panel (Final result)

TESTING REPORT follows "See Note"

See Note

CLINICAL INFORMATION

Tense blisters on urticarial base with pruritus. Presumptive diagnosis is bullous pemphigoid.

Specimen Details

- ; Collected: 3/19/2024; Received: 3/22/2024

DIAGNOSTIC INTERPRETATION

Pemphigoid Antibody Panel monitoring, positive findings supporting the diagnosis of pemphigoid

(See Results, Comments, and Previous and Current Test Results Summary Chart with Graph of ELISA results in the Enhanced Electronic Report/EELR and/or available upon request)

RESULTS

Indirect Immunofluorescence (IIF)

Basement Membrane Zone (BMZ) IgG, IgG4, and IgA Antibodies

IgG: Negative, monkey esophagus substrate
Negative, human split skin substrate

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Patient:
ARUP Accession: 24-079-103960

PCP: Unspecified

IgG4: Detected, titer 1:10 (Borderline), monkey
esophagus substrate
Positive, epidermal localization (roof),
titer 1:20 (H), 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, 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

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)

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

(H) = high/positive

Enzyme-Linked Immunosorbent Assay (ELISA)

Bullous Pemphigoid (BP)180 and BP230 IgG Antibodies

IgG BP180 antibody level: 49 U/mL (H)

Reference Range:
Normal (negative) = Less than 9 U/mL
Increased (H) (positive) = 9 U/mL and greater

IgG BP230 antibody level: 3 U/mL

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Reference Range:

Normal (negative) = Less than 9 U/mL
Increased (H) (positive) = 9 U/mL and greater

(H) = high/positive

U = semiquantitative antibody level in ELISA units

COMMENTS

Specific

The positive IgG4 basement membrane zone antibody reactivity demonstrating epidermal localization (roof) with split skin substrate, also known as salt split skin, by indirect immunofluorescence and the increased IgG BP180 antibody level by ELISA, provide support for the diagnosis of pemphigoid. Previous testing showed findings supporting the diagnosis of pemphigoid and also a normal IgG type VII collagen antibody level by ELISA on one determination. Although IgA basement membrane zone antibodies can be co-expressed with IgG antibodies, no positive IgA basement membrane zone antibody reactivity is detected by indirect immunofluorescence in current or previous testing to indicate this or to support a diagnosis of linear IgA disease or linear IgA/IgG bullous dermatosis. See chart (below) for summary of previous and current basement membrane zone antibody test results; a graph of the ELISA results is available in the Enhanced Electronic Report/EELR and/or available upon request by contacting 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.

Detection, levels, and patterns of diagnostic antibodies may fluctuate with disease manifestations, and IgG BP180 antibody levels correlate with disease activity in some patients with pemphigoid. Clinical correlation is needed, including treatment status, with consideration for continued monitoring of serum antibody profiles by indirect immunofluorescence and antibody levels by ELISAs to aid in assessing disease expression and activity, including response to therapy.

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. IgG4 subclass reactivity by indirect immunofluorescence may be more sensitive than IgG in some patients with pemphigoid and epidermolysis bullosa acquisita. The immunoglobulin class of basement membrane zone antibodies and pattern of antibody localization with split skin substrate (also known as salt split skin)

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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.

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 (also known as bullous pemphigoid antigen 2, BPAG2, or type XVII collagen, COL17) and "BP230" for a 230 kDa bullous pemphigoid antigen (also known as bullous pemphigoid antigen 1, BPAG1). BP180 is a transmembrane component of the basement membrane zone with collagen-like domains; the non-collagenous 16A (NC16A) antigenic domain of BP180 has been identified as a main antigenic target. BP230 is located in the hemidesmosomal plaque of basal cells in the epidermis. Serum levels of IgG BP180 and IgG BP230 antibodies are determined by ELISA, which may be more sensitive than indirect immunofluorescence. Serum levels of IgG BP180 antibodies may correlate with disease activity in pemphigoid, diminishing with treatment response. Up to 7 percent of individuals who do not have pemphigoid, including patients with other immunobullous diseases, have increased levels of IgG BP180 and/or BP230 antibodies by ELISAs.

Patients with pemphigoid may show reactivity to multiple basement membrane zone components in addition to or other than the BP180 and BP230 epitopes in the tested ELISAs. 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 and, potentially, as overlapping basement membrane zone antibody expression in patients with other epithelial antibody-associated disease. Tests that detect antibodies with specificity for other basement membrane zone antigens, including laminin-332, p200 (laminin gamma-1), and alpha6beta4 integrin, may be more sensitive than indirect immunofluorescence but are not currently available, except laminin-332 IgG antibodies in select laboratories. Mucous membrane involvement is predominant in anti-laminin-332 pemphigoid. Recognition of the association of this pemphigoid variant with underlying or developing malignancy (typically solid tumor) in up to one third of cases is critical so appropriate clinical evaluation is conducted. Patients with anti-p200 (laminin gamma-1) pemphigoid tend to be younger than those with bullous pemphigoid and have lesions that clinically resemble both bullous pemphigoid and the inflammatory epidermolysis bullosa acquisita variant that may include mucosal involvement. For those patients with antibodies to alpha6beta4 integrin, alpha6 epitopes primarily are targeted in oral pemphigoid, and beta4 epitopes primarily are targeted in ocular pemphigoid.

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TESTING METHODS

Indirect Immunofluorescence (IIF)

IgG, IgG4, and IgA 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 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. 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, three antibodies on two substrates (IIF X 6)]

Enzyme-Linked Immunosorbent Assays (ELISA)

IgG BP180 and IgG BP230 serum antibody levels determined by U.S. Food and Drug Administration (FDA)-approved ELISAs (Mesacup, MBL BION). [Two ELISAs]

TEST RESULTS SUMMARY CHART

Basement Membrane Zone (BMZ) Antibodies

Serum Number	Date of Specimen	IgG and IgG4 BMZ Titers	IgA BMZ Titers	BP 180	BP 230	Col VII
20-0001	03/26/20	IgG ME Neg IgG SS Neg IgG4 ME 1:5 IgG4 SS Epi 1:20	ME Neg SS Neg	52	4	NA
20-0002	11/15/20	IgG ME Neg IgG SS Neg IgG4 ME 1:10 IgG4 SS Epi 1:20	ME Neg SS Neg	57	4	NA

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21-0003	01/06/21	IgG ME Neg	ME Neg	67	6	NA
		IgG SS Epi,	SS Neg			
			1:10			
		IgG4 ME 1:20				
		IgG4 SS Epi,				
			>1:20			
21-0004	05/20/21	IgG ME Neg	ME Neg	59	5	NA
		IgG SS Epi,	SS Neg			
			1:20			
		IgG4 ME 1:10				
		IgG4 SS Epi,				
			1:20			
21-0005	07/24/21	IgG ME Neg	ME Neg	53	4	2
		IgG SS Epi,	SS Neg			
			1:10			
		IgG4 ME 1:10				
		IgG4 SS Epi,				
			1:20			
24-2515	03/19/24	IgG ME Neg	ME Neg	49	3	NA
		IgG SS Neg	SS Neg			
		IgG4 ME 1:10				
		IgG4 SS Epi,				
			1:20			

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

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Epi = epidermal localization (roof) with split skin substrate (IgG - pemphigoid including bullous pemphigoid, some mucous membrane pemphigoid, and other pemphigoid variants; IgA - linear IgA disease including linear IgA bullous dermatosis and chronic bullous disease of childhood)
Derm = dermal localization (floor) with 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) with 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

Neg = Negative
NA = Not Assayed

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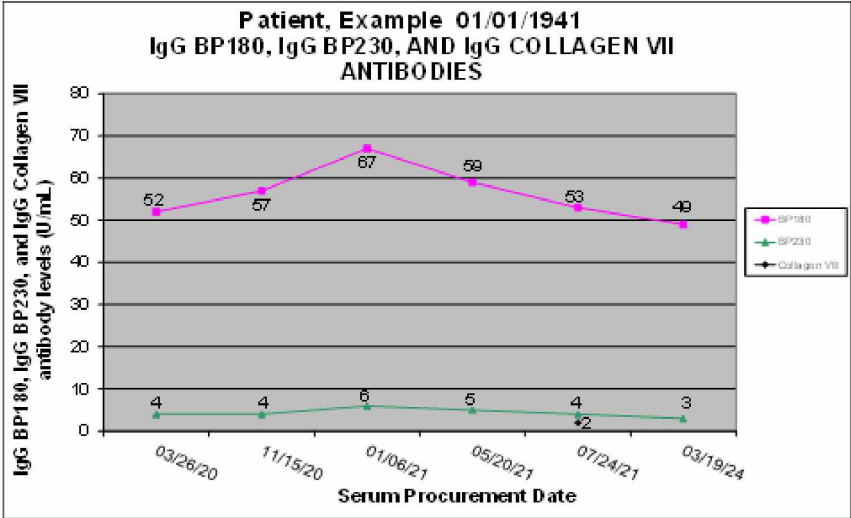
ELISA RESULTS GRAPH (may be found on next page)

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