

500 Chipeta Way, Salt Lake City, Utah 84108-1221

phone: 801-583-2787, toll free: 800-522-2787

Jonathan R. Genzen, MD, PhD, Chief Medical Officer

Patient Age/Sex: 72 years Female

Specimen Collected: 08-Jul-22 10:41

Procedure	Result	Units	Reference Interval
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Bullous Pemphigoid Antigens, IgG	See Note ^{f1}		
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Procedure	Result	Units	Reference Interval
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Bullous Pemphigoid Antigens, IgG	See Note ^{f2}		
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Result Footnote

f1: EER Bullous Pemphigoid Antigens, IgG
Authorized individuals can access the ARUP
Enhanced Report using the following link:

f2: Bullous Pemphigoid Antigens, IgG
CLINICAL INFORMATION
Urticarial plaques, blisters, and pruritus. Presumptive diagnosis is persisting bullous pemphigoid versus
urticarial vasculitis versus contact dermatitis.

Specimen Details

S22-IP0000826 - Serum; Collected: 7/8/2022; Received: 7/11/2022

DIAGNOSTIC INTERPRETATION

Bullous Pemphigoid (BP180 and BP230) IgG Antibodies monitoring, consistent with pemphigoid

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

RESULTS

Enzyme-Linked Immunosorbent Assay (ELISA)

Bullous Pemphigoid (BP)180 and BP230 IgG Antibodies

IgG BP180 antibody level: 64 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

Reference Range:

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

(H) = high/positive

U = antibody level in ELISA units

COMMENTS

* = Abnormal, # = Corrected, C = Critical, f = Result Footnote, H = High, i = Test Information, L = Low, t = Interpretive Text, @ = Performing lab

Unless otherwise indicated, testing performed at:

ARUP Laboratories

500 Chipeta Way, Salt Lake City, UT 84108

Laboratory Director: Jonathan R. Genzen, MD, PhD

ARUP Accession: 22-189-103656

Report Request ID: 16631941

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Result Footnote

f2: Bullous Pemphigoid Antigens, IgG

Specific

The increased IgG BP180 antibody level in this ELISA testing provides support for the diagnosis of pemphigoid. Of note, up to 7 percent of individuals unaffected by pemphigoid, including some with other immunobullous diseases, have increased IgG BP180 and/or IgG BP230 antibody levels, although, generally, those patients who have a relatively high level of IgG BP180 antibodies, as observed in this specimen, are affected with pemphigoid. Previous ELISA testing demonstrated increased IgG BP180 antibody levels. See chart (below) for summary of previous and current 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.

Additional considerations are that co-expression of IgG and IgA basement membrane zone antibodies may develop in patients with pemphigoid, which may have implications for disease severity and treatment approach, or that overlapping antibody expression develops to other basement membrane zone antigens that can be detected by ELISA for IgG type VII collagen antibodies. If indicated to further evaluate the immunopathological profile with respect to basement membrane zone antibodies currently, additional indirect immunofluorescence testing may be performed on this specimen by contacting ARUP Client Services, 1-800-242-2787, option 2, with add-on test request(s) for:

- Basement Membrane Zone (Epithelial) Antibodies, IgG by IIF (ARUP test number 0092056) and
- Basement Membrane Zone (Epithelial) Antibodies, IgA by IIF (ARUP test number 0092057) with or without
- Collagen Type VII Antibody, IgG by ELISA (ARUP test number 2010905).

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

General

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 enzyme-linked immunosorbent assays (ELISA), and 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. In patients with pemphigoid, IgG BP180 and/or IgG BP230 antibody levels, determined by ELISAs, may be more sensitive diagnostic markers than indirect immunofluorescence, and, in patients with epidermolysis bullosa acquisita, the IgG type VII collagen antibody level, determined by ELISA, may be a more sensitive diagnostic marker than indirect immunofluorescence. Patients with pemphigoid or epidermolysis bullosa acquisita may show reactivity to multiple basement membrane zone components in addition to or other than the BP180 and BP230 epitopes and type VII collagen epitopes displayed in the respective ELISAs.

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Result Footnote

f2: Bullous Pemphigoid Antigens, IgG

TESTING METHODS

Enzyme-Linked Immunosorbent Assay (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 Antibodies

Serum Number	Date of Specimen	IgG BMZ Titers	IgA BMZ Titers	BP 180	BP 230	Col VII
21-3011	09/01/21	IgG ME: NA IgG SS: NA	ME: NA SS: NA	96	0	NA
21-4882	11/30/21	IgG ME: NA IgG SS: NA	ME: NA SS: NA	70	1	NA
21-5578	12/15/21	IgG ME: NA IgG SS: NA	ME: NA SS: NA	79	4	NA
22-0496	06/21/22	IgG ME: NA IgG SS: NA	ME: NA SS: NA	64	3	NA

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
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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Result Footnote

f2: Bullous Pemphigoid Antigens, IgG
 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 type VII collagen antibody level (U/mL) by ELISA

NA = Not Assayed

Electronically signed by Kristin M. Leiferman, MD, on 07/11/22 at 6:11 PM.
 Performed At: IMMUNODERMATOLOGY LABORATORY
 417 S. WAKARA WAY, SUITE 2151
 SALT LAKE CITY, UT 84108
 Medical Director: JOHN JOSEPH ZONE, MD
 CLIA Number: 46D0681916

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