

Client: Example Client ABC123
123 Test Drive
Salt Lake City, UT 84108
UNITED STATES

Physician: Doctor, Example

Patient: Patient, Example
DOB 12/31/1998
Gender: Female
Patient Identifiers: 01234567890ABCD, 012345
Visit Number (FIN): 01234567890ABCD
Collection Date: 00/00/0000 00:00

Pemphigoid Gestationis, Complement-Fixing Basement Membrane Antibodies (Herpes Gestationis Factor)
ARUP test code 0092283

EER Herpes Gestationis Factor Ab IgG

See Note
Authorized individuals can access the ARUP
Enhanced Report using the following link:

[Redacted Link]

Herpes Gestationis Factor Ab IgG

See Note
CLINICAL INFORMATION
Pruritic urticarial lesions on abdomen and extremities starting
second trimester of pregnancy.

Specimen Details
[Redacted] - ; Collected: 1/26/2024; Received: 1/29/2024

DIAGNOSTIC INTERPRETATION
Negative for pemphigoid (herpes) gestationis antibodies
(See Results and Comments including further testing
considerations)

RESULTS
Indirect Immunofluorescence (IIF)

Herpes (Pemphigoid) Gestationis Factor (HGF), Complement-Fixing
Basement Membrane Zone (BMZ) Antibodies

Negative, human split skin substrate with
complement
Negative, human split skin substrate without
complement

Reference Range:
Negative - Titer less than 1:2
Borderline - Titer 1:2
Positive (H) - Titer greater than 1:2

Basement Membrane Zone (BMZ) IgG and IgG4 Antibodies
IgG: Negative, human split skin substrate
IgG4: Negative, human split skin substrate

H=High, L=Low, *=Abnormal, C=Critical

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

(H) = high/positive

Enzyme-Linked Immunosorbent Assay (ELISA)

Bullous Pemphigoid (BP)180 IgG Antibodies

IgG BP180 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 = semiquantitative antibody level in ELISA units

COMMENTS

Specific

These negative results for complement-fixing basement membrane zone antibodies on split skin substrate (also known as salt split skin) by indirect immunofluorescence and the normal IgG BP180 antibody level by ELISA are against, but do not rule out, the diagnosis of pemphigoid (herpes) gestationis.

These results do not rule out other immunobullous diseases. In patients with epidermolysis bullosa acquisita and in a subset of patients with bullous lupus erythematosus, the IgG type VII collagen antibody level by ELISA is a diagnostic marker together with dermal (floor) IgG basement membrane zone antibody reactivity with split skin substrate by indirect immunofluorescence. Of note, no specific IgG or IgG4 antibodies localizing to basement membrane zone are detected on split skin substrate by indirect immunofluorescence in this testing, but ELISA may be more sensitive than indirect immunofluorescence. In patients with linear IgA disease, indirect immunofluorescence testing with monkey esophagus substrate and human split skin substrate to detect IgA basement membrane zone antibodies is a sensitive diagnostic marker. In patients with pemphigus, cell surface/intercellular substance antibody testing is a sensitive diagnostic aid. Clinical correlation is needed, including direct immunofluorescence findings on a biopsy specimen and treatment status.

If indicated to further evaluate the immunopathological profile with respect to disease-associated epithelial antibodies, additional testing may be performed on this serum specimen by contacting ARUP Client Services, 1-800-242-2787, option 2, with add-on test request(s) for:

Basement membrane zone antibody-associated diseases (pemphigoid and pemphigoid variants, epidermolysis bullosa acquisita, and linear IgA disease) with the

H=High, L=Low, *=Abnormal, C=Critical

- Basement Membrane Zone Antibody Panel (ARUP test number 3001410);

or

Basement membrane zone and cell surface/intercellular substance antibody-associated diseases (pemphigoid and pemphigoid variants, epidermolysis bullosa acquisita, linear IgA disease, and pemphigus and pemphigus variants) with the

- Immunobullous Disease Antibody Panel (ARUP test number 3001409).

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.

General

Basement membrane zone antibodies with added complement are detected in more than 90 percent of patients with pemphigoid gestationis and without added complement in up to 25 percent of patients. Complement-fixing basement membrane zone antibodies detected by adding fresh serum complement to indirect immunofluorescence testing for herpes gestationis factor also may develop in conditions other than pemphigoid gestationis. IgG4 subclass reactivity by indirect immunofluorescence may be more sensitive than IgG in some patients with pemphigoid but are not the predominant IgG subclass in pemphigoid gestationis, rather it is IgG1.

A major molecular structure in the basement membrane zone to which IgG pemphigoid antibodies bind has 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). 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 for antibodies in patients with bullous pemphigoid and patients with pemphigoid gestationis. Serum levels of IgG BP180 antibodies, determined by ELISA, are in the negative range in normal individuals, and serum levels of IgG BP180 antibodies are increased in most patients with pemphigoid gestationis. IgG BP180 antibody levels by ELISA correlate with disease activity in some patients with bullous pemphigoid but may remain increased in patients with pemphigoid gestationis even with disease remission/resolution.

TESTING METHODS

Indirect Immunofluorescence (IIF)

Herpes (pemphigoid) gestationis factor (HGF), complement-fixing IgG basement membrane zone antibodies with IgG and IgG4 basement membrane zone antibodies

Patient serum is layered on substrate sections of human skin split at the basement membrane zone on microscope slides, neat, 1:2, and progressively in two-fold serum dilutions to a maximum of 1:128, both with and without a fresh source of complement. The substrate sections on slides incubated with serum are then washed and reacted with fluorescein isothiocyanate (FITC)-conjugated antibody to C3. FITC-conjugated anti-IgG also is tested on the human split skin substrate sections with serum two-fold dilutions from 1:5 to maximum of 1:40,960. The limiting-dilution, end-point titers and patterns of staining on split skin substrate are reported. Additionally, FITC-conjugated anti-IgG4 is tested to increase sensitivity (maximum serum dilution of 1:20). This indirect immunofluorescence testing, including complement fixation, 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

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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, one antibody on one substrate, with and without complement, and two antibodies on one substrate (IIF X 4)]

Enzyme-Linked Immunosorbent Assay (ELISA)

IgG BP180 serum antibody level determined by U.S. Food and Drug Administration (FDA)-approved ELISA (Mesacup, MBL BION). (One ELISA)

Electronically signed by [REDACTED] on 02/07/24 at 1:26 PM.
Performed At: IMMUNODERMATOLOGY LABORATORY
417 S. WAKARA WAY, SUITE 2151
SALT LAKE CITY, UT 84108
Medical Director: KRISTIN M. LEIFERMAN, MD
CLIA Number: 46D0681916

VERIFIED/REPORTED DATES				
Procedure	Accession	Collected	Received	Verified/Reported
EER Herpes Gestationis Factor Ab IgG	24-026-122741	00/00/0000 00:00	00/00/0000 00:00	00/00/0000 00:00
Herpes Gestationis Factor Ab IgG	24-026-122741	00/00/0000 00:00	00/00/0000 00:00	00/00/0000 00:00

END OF CHART

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