ARUP LABORATORIES | aruplab.com
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 REPORT

Patient Age/Sex: 27 years Female

Specimen Collected: 21-Jun-22 15:42

Herpes Gestationis Factor Antibody IgG **|Received: 22-Jun-22 09:57** **Report/Verified: 22-Jun-22 11:13**

Procedure	Result	Units	Reference Interval
Herpes Gestationis Factor Ab IgG	See Note [f1]		

Result Footnote

f1: Herpes Gestationis Factor Ab IgG
CLINICAL INFORMATION
Pruritic, urticarial, and blistering lesions onset late second trimester of pregnancy. Presumptive diagnosis is pemphigoid gestationis versus other dermatosis of pregnancy.

Specimen Details
S22-IP0000502 - Serum; Collected: 6/21/2022; Received: 6/22/2022

DIAGNOSTIC INTERPRETATION
Consistent with pemphigoid (herpes) gestationis; positive for herpes gestationis factor

(See Results and Comments)

RESULTS
Indirect Immunofluorescence (IIF)

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

Positive, epidermal pattern (roof), titer 1:16,
human split skin substrate with complement
Positive, epidermal pattern (roof), titer 1:4, 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 Antibodies

IgG: Positive, epidermal pattern (roof), titer 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 BMZ antibodies = pemphigoid
(including pemphigoid gestationis, bullous
pemphigoid, mucous membrane pemphigoid)

Dermal (floor) IgG BMZ antibodies = epidermolysis
bullosa acquisita or bullous lupus erythematosus

Result Footnote

f1: Herpes Gestationis Factor Ab IgG
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: 63 U/mL (H)

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

COMMENTS

Specific

The positive complement-fixing basement membrane zone antibodies (so-called herpes gestationis factor) detected in this serum specimen together with an increased IgG BP180 antibody level by ELISA, support the diagnosis of pemphigoid (herpes) gestationis. Approximately 25 percent of patients with pemphigoid gestationis have IgG basement membrane zone antibodies, as observed in this testing. This pattern of immunological reactivity can be observed in other types of pemphigoid. Clinical correlation is needed including with direct immunofluorescence findings on a biopsy specimen showing characteristic strong linear C3 basement membrane zone localization with weaker or absent linear IgG basement membrane zone antibody reactivity.

Of note, positive celiac disease serologies have been identified in a subset of patients with pemphigoid gestationis. If indicated to further evaluate the immunopathological profile, contact ARUP Client Services at 1-800-242-2787, option 2, with add-on test request(s) for:
- Celiac Disease Reflexive Cascade (ARUP test number 2008114), and
- Epidermal Transglutaminase (eTG/TG3) Antibody, IgA by ELISA (ARUP test number 2010902).

IgG BP180 antibody levels correlate with disease activity in some patients with bullous pemphigoid but may remain increased in patients with pemphigoid gestationis even with disease remission/resolution. Detection, levels, and patterns of diagnostic antibodies may fluctuate with disease manifestations. Monitoring serum antibody profiles by indirect immunofluorescence and antibody levels by ELISAs may aid in assessing disease expression and activity, particularly for persisting, progressing, or changing disease.

General

Basement membrane zone antibodies without added complement are detected in up to 25 percent of patients with pemphigoid gestationis and with added complement in more than 90 percent. Complement-fixing basement membrane zone antibodies detected by adding fresh serum complement to indirect immunofluorescence testing for herpes gestationis factor may be expressed in conditions other than pemphigoid gestationis.

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

Result Footnote

f1: Herpes Gestationis Factor Ab IgG
pemphigoid antigen 2, BPAG2, or type XVII collagen, COL17). BP180 is a transmembrane component of the basement membrane zone with collagen-like domains and is a principal antigenic target of basement membrane zone antibodies both in patients with bullous pemphigoid and patients with pemphigoid gestationis. Serum levels of IgG BP180 antibodies, determined by enzyme-linked immunosorbent assay (ELISA), 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 basement membrane zone antibodies

Patient serum is layered on substrate sections of human skin split at the basement membrane zone on microscope slides, at 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 in 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. 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 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 one antibody on one substrate (IIF X 3), with two limiting-dilution, end-point titers (antibody titer X 2)]

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 Kristin M. Leiferman, MD, on 06/22/22 at 11:09 AM.
Performed At: IMMUNODERMATOLOGY LABORATORY
417 S. WAKARA WAY, SUITE 2151
SALT LAKE CITY, UT 84108
Medical Director: JOHN JOSEPH ZONE, MD
CLIA Number: 46D0681916