

Client: Example Client ABC123
123 Test Drive

UNITED STATES

Physician: Doctor, Example

Patient: Patient, Example

DOB

Sex:

Patient Identifiers: 01234567890ABCD, 012345

Visit Number (FIN): 01234567890ABCD

Collection Date: 01/01/2017 12:34

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

ARUP test code 0092283

Herpes Gestationis Factor Ab IgG

See Note

CLINICAL INFORMATION

Pruritic urticarial lesions on abdomen and extremities starting second trimester of pregnancy.

Specimen Details

S22-IP0000523 - Serum; Collected: ; Received:

DIAGNOSTIC INTERPRETATION

Negative for pemphigoid (herpes) gestationis factor/complement-fixing basement membrane zone antibodies

(See Results and Comments)

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 Antibodies

IgG: 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 BMZ antibodies = pemphigoid (including pemphigoid gestationis, bullous pemphigoid, mucous membrane pemphigoid)

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

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

Unless otherwise indicated, testing performed at:

ARUP LABORATORIES | 800-522-2787 | aruplab.com
500 Chipeta Way, Salt Lake City, UT 84108-1221
Jonathan R. Genzen, MD, PhD, Laboratory Director

Patient: Patient, Example
ARUP Accession: 22-173-114892
Patient Identifiers: 01234567890ABCD, 012345
Visit Number (FIN): 01234567890ABCD
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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 = 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 pattern IgG basement membrane zone antibody reactivity on split skin substrate by indirect immunofluorescence. In patients with linear IgA disease, indirect immunofluorescence testing on monkey esophagus substrate and human split skin substrate to detect IgA basement membrane zone antibodies is a sensitive diagnostic aid. In patients with pemphigus, cell surface antibody testing is a sensitive diagnostic aid. Clinical correlation is needed including with treatment status and with direct immunofluorescence findings on a biopsy specimen which demonstrates strong linear C3 basement membrane zone localization with weaker or absent linear IgG basement membrane zone antibody reactivity in patients with the disease.

If indicated to further evaluate the immunopathological profile, 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

- Basement Membrane Zone (Epithelial) Antibodies, IgG by IIF (ARUP test number 0092056),
- Basement Membrane Zone (Epithelial) Antibodies, IgA by IIF (ARUP test number 0092057),
- Bullous Pemphigoid (BP180 and BP230) Antibodies, IgG by ELISAs (ARUP test number 0092566), and/or
- Collagen Type VII Antibody, IgG by ELISA (ARUP test number 2010905);

or

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

- Basement Membrane Zone and Cell Surface (Epithelial) Antibodies, IgG and IgA by IIF (ARUP test number

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- 0090299), and
- Bullous Pemphigoid (BP180 and BP230) Antibodies, IgG by ELISAs (ARUP test number 0092566),
- Collagen Type VII Antibody, IgG by ELISA (ARUP test number 2010905), and
- Desmoglein 1 and Desmoglein 3 (Pemphigus) Antibodies, IgG by ELISA (ARUP test number 0090649).

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 also may develop 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 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 both bullous pemphigoid and 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 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)]

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

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ELISA]

Electronically signed by _____, MD, on _____
at _____
Performed At: _____

Medical Director: _____, MD
CLIA Number: _____

VERIFIED/REPORTED DATES

Procedure	Accession	Collected	Received	Verified/Reported
Herpes Gestationis Factor Ab IgG	22-173-114892			

END OF CHART

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