

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

Physician: Doctor, Example

Patient: Patient, Example

DOB 12/31/1976 **Gender:** Female

Patient Identifiers: 01234567890ABCD, 012345

Visit Number (FIN): 01234567890ABCD **Collection Date:** 00/00/0000 00:00

Collagen Type VII Antibody, IgG by ELISA

ARUP test code 2010905

Collagen Type VII Antibody IgG by ELISA

See Note

CLINICAL INFORMATION

Skin fragility, tense blisters, vesicles, erosions, and milia. Presumptive diagnosis is epidermolysis bullosa acquisita versus porphyria cutaneous tarda.

Specimen Details S22-IP0000497 - Serum; Collected: 6/21/2022; Received: 6/22/2022

DIAGNOSTIC INTERPRETATION

Increased IgG type VII collagen antibody level by ELISA and concurrent indirect immunofluorescence testing demonstrates positive IgG, including IgG4, basement membrane zone antibodies with dermal localization (floor) on split skin substrate; consistent with subepidermal immunobullous disease, including epidermolysis bullosa acquisita or bullous lupus erythematosus

(See Results, Comments, separate concurrent Basement Membrane Zone (Epithelial) Antibodies, IgG by IIF testing report with additional findings and comments, and Basement Membrane Zone Antibody Test Results Summary Chart with concurrent findings)

RESULTS

Enzyme-Linked Immunosorbent Assay (ELISA)

Type VII Collagen IgG Antibodies

IgG type VII collagen antibody level: 88 U/mL (H)

Reference Range:

Normal (negative) = Less than 7 U/mL Slightly increased (H) (positive) = 7-8 U/mL Increased (H) (positive) = 9 U/mL and greater

COMMENTS

Specific

The IgG type VII collagen antibody level is increased in this ELISA testing, which, combined with the dermal (floor) IgG antibody reactivity on split skin substrate, also known as salt split skin, by indirect immunofluorescence in concurrent testing (separate report with additional comments), supports the diagnosis of epidermolysis bullosa acquisita and bullous lupus erythematosus. See chart at end of report (below) for summary of concurrent basement membrane zone antibody test results.

Patients with inflammatory bowel disease, including Crohn

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



disease and ulcerative colitis, with and without mucocutaneous manifestations of epidermolysis bullosa acquisita or bullous lupus erythematosus, also may demonstrate increased antibodies to type VII collagen. As noted in the concurrent report, two subsets of pemphigoid, namely, anti-laminin-332 and anti-p200 (laminin gamma-1) pemphigoid, demonstrate IgG basement membrane zone antibody reactivity with the dermal side of the split skin substrate by indirect immunofluorescence, although these two pemphigoid subsets do not characteristically demonstrate increased levels of IgG type VII collagen antibodies, as observed in this testing. Therefore, although the overall immunopathological profile is consistent with epidermolysis bullosa acquisita or, less commonly, with bullous lupus erythematosus, the findings do not entirely rule out anti-laminin-332 pemphigoid or anti-p200 (laminin gamma-1) pemphigoid with increased IgG type VII collagen antibodies associated with another condition.

Other than this IgG type VII collagen antibody determination by ELISA, the disorders with dermal pattern IgG basement membrane zone reactivity cannot be readily distinguished based on currently available diagnostic laboratory techniques. It is important to note that up to one third of patients with anti-laminin-332 pemphigoid have or will develop an associated malignancy. Therefore, clinical correlation is needed with further clinical evaluation as indicated. Correlation with direct immunofluorescence findings on a biopsy specimen also is recommended. Monitoring serum antibody profiles by indirect immunofluorescence and antibody levels by ELISAs may aid in assessing disease expression and activity, including response to therapy.

General

Type VII collagen is a component of anchoring fibrils within epithelial basement membrane zone (skin and mucous membranes), and patients with epidermolysis bullosa acquisita characteristically develop IgG antibodies to type VII collagen. An increased serum IgG type VII collagen antibody level by ELISA provides support for the diagnosis of epidermolysis bullosa acquisita and also a subset of bullous lupus erythematosus together with dermal localization (floor) of IgG basement membrane zone antibodies on split skin substrate by indirect immunofluorescence. Patients with inflammatory bowel disease, including Crohn disease and ulcerative colitis, with and without mucocutaneous manifestations of epidermolysis bullosa acquisita, may demonstrate increased levels of antibodies to type VII collagen. The major epitopes for antibody reactivity reside in the non-collagenous amino-terminal domain, NC1, with minor epitopes in the non-collagenous carboxy-terminal domain, NC2, of the three identical alpha chains that comprise type VII collagen. The tested ELISA contains combined purified recombinant antigens from both NC1 and NC2 for detection of IgG antibodies. Serum antibody levels above the reference range threshold of 6 U/mL may correlate with disease activity. Patients with epidermolysis bullosa acquisita or bullous lupus erythematosus may develop antibodies to basement membrane zone antigens in addition to or other than the type VII collagen epitopes displayed in this ELISA, and patients with other epithelial antibody-associated disorders may develop overlapping basement membrane zone antibody expression with an increased level of IgG type VII collagen antibodies.

TESTING METHODS
Enzyme-Linked Immunosorbent Assay (ELISA)

IgG type VII collagen serum antibody level determined by ELISA (Mesacup, MBL International). The performance characteristics of this ELISA testing were determined by the Immunodermatology Laboratory at the University of Utah. The testing has not been cleared or approved by the FDA (US Food and Drug

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Patient: Patient, Example
ARUP Accession: 22-172-113406
Patient Identifiers: 01234567890ABCD, 012345
Visit Number (FIN): 01234567890ABCD
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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. [One ELISA]

TEST RESULTS SUMMARY CHART Basement Membrane Zone Antibodies

Serum Number	Date of Specimen	IgG BMZ		d IgG4 ters	Ig/ BM2	A Z Titers	BP 180	BP 230	Col VII
22-0497	06/21/22	IgG IgG IgG4 IgG4	SS ME	NA NA		NA NA	NA	NA	88
22-0499	06/21/22	IğG	SS	1:5120 Derm, 1:2560 >1:20		NA NA	NA	NA	NA
				Derm, >1:20					

Chart Key:

IgG BMZ = IgG basement membrane zone (BMZ) antibodies by

indirect immunofluorescence

= IgG4 basement membrane zone (BMZ) antibodies

by indirect immunofluorescence
= IgA basement membrane zone (BMZ) antibodies by
indirect immunofluorescence IgA BMZ

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

= epidermal localization (roof) on split skin

Epi = epidermal localization (roof) on 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) 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) acquisită)

Comb = combined epidermal-dermal localization (roof and floor) on split skin substrate (IgG -pemphigoid and pemphigoid variants; IgA - linear IgA disease)

 $\tt BP180 = IgG \ BP180$ antibody level (U/mL) by ELISA $\tt BP230 = IgG \ BP230$ antibody level (U/mL) by ELISA Col VII = IgG Collagen VII antibody level (U/mL) by

NA = Not Assayed

Electronically signed by

Performed At: IMMUNODERMATOLOGY LABORATORY 417 S. WAKARA WAY, SUITE 2151 SALT LAKE CITY, UT 84108

Medical Director: JOHN JOSEPH ZONE, MD

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CLIA Number: 46D0681916

EER Collagen Type VII Ab IgG, by ELISA

See Note

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

VERIFIED/REPORTED DATES									
Procedure	Accession	Collected	Received	Verified/Reported					
Collagen Type VII Antibody IgG by ELISA	22-172-113406	00/00/0000 00:00	00/00/0000 00:00	00/00/0000 00:00					
EER Collagen Type VII Ab IgG, by ELISA	22-172-113406	00/00/0000 00:00	00/00/0000 00:00	00/00/0000 00:00					

END OF CHART

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