Collagen Type VII Antibody, IgG by ELISA

ARUP LABORATORIES

Patient: [redacted]
DOB: [redacted] Age: [redacted] Sex: [redacted]
Patient Identifiers: [redacted]

Visit Number (FIN): [redacted]

Client: [redacted]

Physician: [redacted]

ARUP Test Code: 2010905

Collection Date: 06/21/2022
Received in lab: 06/22/2022
Completion Date: 06/22/2022

Immunodermatology Serum Test Report Navigation Guide

The Immunodermatology TESTING REPORT from the University of Utah follows "See Note" and is arranged as outlined below on the following pages:

CLINICAL INFORMATION
This content is provided by the ordering clinician and includes the reason for testing.

Specimen Details
This includes specimen identification with collected and received dates.

DIAGNOSTIC INTERPRETATION
This is a synopsis of key findings from the testing and their diagnostic relevance.

RESULTS
This section reports the discrete finding and value of each test component, along with the reference range.

COMMENTS
Specific

These comments provide an explanation of the test results as they relate to clinical considerations, and include reference to any concurrent and/or previous testing.

General

These comments summarize fundamental information about the test(s) and the component(s) assessed to aid in interpretation of their clinical applicability.

TESTING METHODS
The section lists the procedures performed, the test source(s), and the applicable laboratory developed test disclaimer(s).

TEST RESULTS SUMMARY CHART
A chart tabulating results of tests ordered for the patient by the same client is included if previous and/or concurrent testing has been performed.

ELISA RESULTS GRAPH
A graph of ELISA results also is included if previous and/or concurrent testing has been performed; the graph may be found on a subsequent page.

For testing algorithm and additional information, refer to:
arupconsult.com/content/immunobullous-skin-diseases-screening

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Chart continues on following page(s)

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UNIVERSITY OF UTAH

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IMMUNODERMATOLOGY LABORATORY REPORT

[redacted]

Submitter

ARUP Sendouts

Collagen Type VII Antibody, IgG by ELISA (Final result)

TESTING REPORT follows "See Note"
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

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[redacted]
PCP: Unspecified

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 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

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PCP: Unspecified

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 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 and IgG4 BMZ Titers	IgA BMZ Titers	BP 180	BP 230	Col VII
22-0497	06/21/22	IgG ME NA IgG SS NA IgG4 ME NA IgG4 SS NA	ME NA SS NA	NA	NA	88
22-0499	06/21/22	IgG ME 1:5120 IgG SS Derm, 1:2560	ME NA SS NA	NA	NA	NA

[redacted]
PCP: Unspecified

IgG4 ME>1:20
IgG4 SS Derm, >1:20

Chart Key:

IgG BMZ = IgG basement membrane zone (BMZ) antibodies by indirect immunofluorescence
IgG4 BMZ = IgG4 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 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 Collagen VII antibody level (U/mL) by ELISA

NA = Not Assayed

Electronically signed by [redacted], MD, on 06/22/22 at 12:51 PM.

Resulting Laboratory

[redacted]
PCP: Unspecified

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