Envoplakin 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: 3016533 Collection Date: 11/20/2024 Received in lab: 11/22/2024 Completion Date: 11/22/2024

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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IMMUNODERMATOLOGY LABORATORY REPORT

[redacted]

Submitter

ARUP Sendouts

Envoplakin Antibody, IgG by ELISA (Final result)

TESTING REPORT follows "See Note"
See Note

CLINICAL INFORMATION
Mucosal involvement with extensive erosions and targetoid, urticarial, and scaly skin lesions. Presumptive diagnosis is pemphigus, including paraneoplastic pemphigus, versus drug reaction, Stevens-Johnson syndrome.

Specimen Details
[redacted] - ; Collected: 11/20/2024; Received: 11/22/2024

DIAGNOSTIC INTERPRETATION

Increased IgG envoplakin antibody level by ELISA supporting a diagnosis of paraneoplastic pemphigus/paraneoplastic autoimmune multiorgan syndrome

(See Results and Comments including recommendations for correlating with other testing)

RESULTS
Enzyme-Linked Immunosorbent Assay (ELISA)

Envoplakin IgG Antibodies

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[redacted]
PCP: Unspecified

IgG envoplakin antibody level: 1.51 ratio U/mL (H)

Reference Range:
Normal (negative) = Less than 1.00 ratio U/mL
Increased (H) (positive) = 1.00 ratio U/mL and greater

(H) = high/positive
U = antibody level in ELISA units

COMMENTS

Specific

The increased level of IgG envoplakin antibodies in this testing supports the diagnosis of paraneoplastic pemphigus (PNP), also known as paraneoplastic autoimmune multiorgan syndrome (PAMS). Envoplakin antibodies may be increased in patients with other diseases; therefore, clinical correlation is needed, including with direct immunofluorescence test findings on a biopsy specimen, histopathological examination of formalin-fixed tissue, and other serum epithelial antibodies.

Notably, various serum epithelial antibodies may be found in paraneoplastic pemphigus and other paraneoplastic presentations by various tests with differing sensitivities. IgG paraneoplastic pemphigus antibody reactivity with rat bladder, mouse bladder, mouse heart, and mouse liver substrates by indirect immunofluorescence may provide confirmatory broad-based testing for paraneoplastic pemphigus antibodies, and testing for epithelial cell surface/intercellular substance and basement membrane zone antibodies by indirect immunofluorescence and ELISAs can be helpful to expand the antibody profile for other potential epithelial targets.

To further evaluate the immunopathological profile, additional testing may be performed on this serum specimen, as clinically indicated, by contacting ARUP Client Services at 1-800-242-2787, option 2, with add-on test request(s) for:
- Paraneoplastic Pemphigus (Paraneoplastic Autoimmune Multiorgan Syndrome) Screening Antibodies by IIF (rodent substrates including rat bladder) (ARUP test number 0092107),
- Pemphigus Antibody Panel, IgG (ARUP test number 0090650),
- Pemphigus Antibodies, IgA by IIF (ARUP test number 0092106), and
- Basement Membrane Zone Antibody Panel (ARUP test number 3001410);

Or specific antibodies that may develop to other epithelial antigens in paraneoplastic pemphigus,
- Desmoglein 1 and Desmoglein 3 (Pemphigus) Antibodies, IgG by ELISA (ARUP test number 0090649, and

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PCP: Unspecified

- Bullous Pemphigoid (BP180 and BP230) Antibodies, IgG by ELISA (ARUP test number 0092566);

Or the test panels that include all of the above,

- Paraneoplastic Pemphigus (Paraneoplastic Autoimmune Multiorgan Syndrome) Screening Antibodies by IIF (rodent substrates including rat bladder) (ARUP test number 0092107), and
- Immunobullous Disease Antibody Panel (ARUP test number 3001409).

IgG envoplakin antibody levels may correlate with disease activity in patients with PNP/PAMS; monitoring antibody profiles by indirect immunofluorescence as well as antibody levels by ELISAs may aid in assessing disease expression and activity, including response to therapy.

If it would be helpful to discuss the patient case with this report, including request for additional testing on this serum specimen, 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

IgG envoplakin antibodies develop in many patients with paraneoplastic pemphigus (PNP), also known as paraneoplastic autoimmune multiorgan syndrome (PAMS), and rarely in patients with other immunobullous diseases. This ELISA identifies IgG antibodies to the N-terminal recombinant fragment of envoplakin. Specificity of the test has been reported to be high at 96-99 percent and sensitivity of the test more variably reported at 55-86 percent. Patients with PNP/PAMS develop serum antibodies to multiple epithelia (simple, columnar, transitional) with several possible epithelial targets, including plakins (envoplakin, periplakin, desmoplakin I, desmoplakin II, epiplakin, plectin, BP230), also cadherins (desmoglein 1, desmoglein 3; desmocollin 1, desmocollin 2, desmocollin 3), alpha-2-macroglobulin-like-1 (A2ML1), laminin 332, and/or BP180. Envoplakin and periplakin are principal antigenic targets in the disease, and, based on the high specificity, an increased envoplakin antibody level is a diagnostic marker for PNP/PAMS. ELISA may be more sensitive than indirect immunofluorescence testing for detecting antibodies, especially low levels, but indirect immunofluorescence testing with rodent substrates demonstrates antibodies to a broader range of epithelial targets than the envoplakin epitopes displayed in this tested ELISA.

Paraneoplastic pemphigus/paraneoplastic autoimmune multiorgan syndrome may affect all ages and develops as a severe mucocutaneous blistering and erosive disease in association with malignancies, most often hematologic (lymphoma, leukemia) and sarcoma. It also may develop in association with benign neoplasia, especially Castleman disease, which is the most frequent association in children and adolescents. Antibodies

[redacted]
PCP: Unspecified

targeting the various types of epithelia can lead to involvement of various organs and tissues, for example, eyes, lungs, gastrointestinal tract, kidney, and thyroid and is the basis of the name, paraneoplastic autoimmune multiorgan syndrome. IgG envoplakin antibody levels correlate with extent of mucocutaneous paraneoplastic pemphigus disease expression. After successful tumor therapy, envoplakin antibody levels decrease significantly.

For positive/increased ELISA results without known malignancy, perform aggressive evaluation and monitoring for malignancy. For negative results, correlate with histopathological examination of formalin-fixed tissue in addition to direct immunofluorescence testing on a biopsy specimen and epithelial antibodies in serum characteristic of other immunobullous diseases with further clinical evaluation as indicated. Negative IgG envoplakin ELISA testing results do not rule out paraneoplastic/malignancy-associated disease. Other known paraneoplastic epithelial autoantibody associations include nonclassical, intercellular IgG/IgA dermatosis and anti-laminin 332 pemphigoid.

TESTING METHODS
Enzyme-Linked Immunosorbent Assay (ELISA)

IgG envoplakin serum antibody level determined by ELISA (Euroimmun US, Inc.). The performance characteristics of this test have been validated by the University of Utah Immunodermatology Laboratory and meet specifications required by the Clinical Laboratory Improvement Amendment of 1988 (CLIA). The test has CE Mark certification as an in vitro diagnostic in the European Union. The testing has not been cleared or approved by the US Food and Drug Administration (FDA). The testing was performed in a CLIA-certified laboratory and is intended for clinical purposes. The results are not intended to be used as the sole means for clinical diagnosis or patient management decisions. [One ELISA]

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Resulting Laboratory

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