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Jonathan R. Genzen, MD, PhD, Chief Medical Officer

**PATIENT REPORT**

Patient Age/Sex: 53 years Female

**Specimen Collected: 30-May-23 13:48**

**Envoplakin Antibody, IgG by ELISA | Received: 30-May-23 13:58 Report/Verified: 30-May-23 14:26**

| Procedure | Result | Units | Reference Interval |
|---|---|---|---|
| Envoplakin Antibody,IgG by ELISA | See Note [f1] | | |

**Envoplakin Antibody, IgG by ELISA | Received: 30-May-23 13:54 Report/Verified: 30-May-23 14:35**

| Procedure | Result | Units | Reference Interval |
|---|---|---|---|
| EER Envoplakin Antibody,IgG by ELISA | See Note [f2] | | |

## Result Footnote

f1: Envoplakin Antibody, IgG by ELISA
CLINICAL INFORMATION
No clinical information provided.
Specimen Details
S23-IP0000276 - Serum; Collected: 5/30/2023; Received: 5/30/2023

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DIAGNOSTIC INTERPRETATION
Testing interface
(See Results and Comments)

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RESULTS
Enzyme-Linked Immunosorbent Assay (ELISA)

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Envoplakin IgG Antibodies
IgG envoplakin antibody level: 15 ratio U/mL (H)
Reference Range:
Normal (negative) = Less than 1.0 ratio U/mL
Increased (H) (positive) = 1.0 ratio U/mL and greater
(H) = high/positive
U = antibody level in ELISA units

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COMMENTS
Specific

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The increased level of IgG envoplakin antibodies in this testing supports the diagnosis of paraneoplastic pemphigus. Clinical correlation is needed, including with direct immunofluorescence 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 antibodies to 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 and basement membrane zone antibodies can be helpful to evaluate for other immunobullous diseases. If indicated to further evaluate the immunopathological profile, additional testing may be performed on this serum specimen by contacting the Immunodermatology Laboratory with add-on test request(s) for:

- Paraneoplastic Pemphigus Antibody Screen (indirect immunofluorescence on rodent substrates); and
- IgG Pemphigus Antibody Panel; and/or
- IgA Pemphigus (IgA cell surface) Antibody; and/or
- Basement Membrane Zone Antibody Panel.

Or the test panels that include all of the above:

- Paraneoplastic Pemphigus Antibody Screening Panel and
- Immunobullous Disease Antibody Panel.

IgG envoplakin antibody levels may correlate with disease activity in paraneoplastic pemphigus;

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*=Abnormal, #=Corrected, C=Critical, f=Result Footnote, H-High, i-Test Information, L-Low, t-Interpretive Text, @=Performing lab

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## Result Footnote

f1: Envoplakin Antibody, IgG by ELISA
monitoring antibody profiles by indirect immunofluorescence and 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, contact the University of Utah Immunodermatology Laboratory at (US) 1-801-581-7139 or 1-866-266-5699.
General
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IgG envoplakin antibodies develop in patients with paraneoplastic pemphigus (paraneoplastic autoimmune multiorgan syndrome) 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 at 96-99 percent and sensitivity at 80-86 percent. Patients with paraneoplastic pemphigus (paraneoplastic autoimmune multiorgan syndrome) 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 paraneoplastic pemphigus (paraneoplastic autoimmune multiorgan syndrome).

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 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 ELISA testing results without known malignancy, perform aggressive evaluation for malignancy. For negative results, correlate with histopathological examination of formalin-fixed tissue in addition to direct immunofluorescence on a biopsy specimen and epithelial antibodies in serum characteristic of other immunobullous diseases. Negative IgG envoplakin ELISA testing results do not rule out paraneoplastic/malignancy-associated disease.

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TESTING METHODS
Enzyme-Linked Immunosorbent Assay (ELISA)
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IgG envoplakin serum antibody level determined by ELISA (Euroimmun US, Inc.). The performance characteristics of this test have been determined and validated by the Immunodermatology Laboratory at the University of Utah. The U.S. Food and Drug Administration (FDA) has not approved or cleared this test; however, FDA clearance or approval currently may not be required for this testing performed in a CLIA-certified laboratory (Clinical Laboratory Improvement Amendments) and intended for clinical purposes.[One ELISA, 83516]
Electronically signed by Marjorie Allen, on 05/30/23 at 2:15 PM.

Performed At: IMMUNODERMATOLOGY LABORATORY
417 S. WAKARA WAY, SUITE 2151
SALT LAKE CITY, UT 84108
Medical Director: KRISTIN M. LEIFERMAN, MD
CLIA Number: 46D0681916

f2: EER Envoplakin Antibody, IgG by ELISA
Authorized individuals can access the ARUP Enhanced Report using the following link:

## Result Footnote

f2: EER Envoplakin Antibody, IgG by ELISA

[redacted]

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