

**NEW TEST**

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**Paraneoplastic Pemphigus (Paraneoplastic Autoimmune Multiorgan Syndrome) Expanded Antibody Panel by IIF With ELISA**

3016534, PNP PLUS

**Specimen Requirements:**

**Patient Preparation:**

**Collect:** Plain red or serum separator tube (SST).

**Specimen Preparation:** Transfer 2 mL serum to an ARUP standard transport tube. (Min: 0.5 mL)

**Transport Temperature:** Refrigerated

**Unacceptable Conditions:** Hemolyzed or lipemic specimens. Plasma.

**Remarks:**

**Stability:** Ambient: 1 week; Refrigerated: 2 weeks; Frozen: Indefinitely

**Methodology:** Semi-Quantitative Indirect Immunofluorescence (IIF)/Semi-Quantitative Enzyme-Linked Immunosorbent Assay (ELISA)

**Performed:** Varies

**Reported:** 7-14 days

**Note:** The methodology is indirect immunofluorescence (IIF) of patient serum with rodent substrates including rat bladder, mouse bladder, mouse heart, and mouse liver to detect characteristic IgG antibody reactivity: simple columnar epithelial cell surface and basement membrane zone in bladders, intercalated discs in heart, and portal tracts in liver, and enzyme-linked immunosorbent assay (ELISA) to detect IgG antibodies to envoplakin. Monkey esophagus substrate is included if other concurrent IIF testing does not. This test should be distinguished from antibody testing of cerebral spinal fluid (CSF) for paraneoplastic neurologic syndromes; 3004510, 3004512, 3004517 are different tests. Paraneoplastic pemphigus (PNP), also known as paraneoplastic autoimmune multiorgan syndrome (PAMS), is a rare paraneoplastic disease that affects patients of all ages, is associated with lymphoproliferative disorders/malignancies and demonstrates clinical features of severe pemphigus with a high mortality rate. Patients with PNP/PAMS develop serum antibodies to multiple epithelia (simple, columnar, transitional)

with several possible epithelial antigen targets. Envoplakin is one of several possible epithelial targets, albeit a major one, and IgG envoplakin antibody levels correlate with extent of mucocutaneous disease in patients with PNP/PAMS.

CPT Codes: 88346; 88350 x3; 83516

New York DOH Approval Status: This test is New York DOH approved.

Interpretive Data:

Refer to report

Reference Interval:

By report

**HOTLINE NOTE: Refer to the Hotline Test Mix for interface build information.**