

Overview**Useful For**

Diagnosis of paraneoplastic pemphigus/paraneoplastic autoimmune multiorgan syndrome (PNP/PAMS) in the setting of erosive or lichenoid mucocutaneous disease

Method Name

Indirect Immunofluorescence

NY State Available

Yes

Specimen**Specimen Type**

Serum

Specimen Required**Container/Tube:**

Preferred: Red top

Acceptable: Serum gel

Specimen Volume: 2 mL

Specimen Minimum Volume

0.5 mL

Reject Due To

| | |
|-----------------|--------|
| Gross hemolysis | Reject |
| Gross lipemia | Reject |
| Gross icterus | Reject |

Specimen Stability Information

| Specimen Type | Temperature | Time | Special Container |
|---------------|--------------------------|---------|-------------------|
| Serum | Refrigerated (preferred) | 14 days | |
| | Frozen | 30 days | |
| | Ambient | 14 days | |

Clinical and Interpretive**Clinical Information**

Paraneoplastic pemphigus (PNP; also paraneoplastic autoimmune multiorgan syndrome: PAMS, to denote the systemic nature of the syndrome) is an autoimmune mucocutaneous blistering disease affecting adults or rarely children that generally heralds the presence of an underlying malignancy.

PNP/PAMS can be defined and identified by a combination of the following features: 1) painful stomatitis and a polymorphous cutaneous eruption with lesions that may be blistering, lichenoid, erythema multiforme-like or morbilliform; 2) variable histopathologic findings, including acantholysis, lichenoid, or interface change; 3) variable direct immunofluorescence findings from a perilesional biopsy, often demonstrating deposition of IgG and complement in the epidermal intercellular spaces, granular/linear complement deposition along the epidermal basement membrane zone, and/or a lichenoid tissue reaction; 4) indirect immunofluorescence evidence of cell surface deposition on monkey esophagus and/or rat bladder epithelium, 5) ELISA evidence of serum autoantibodies against desmogleins 1 or 3, and possibly against bullous pemphigoid (BP) 180 and 230 antigens. The incidence of the disease is unknown but it is less common than pemphigus vulgaris (PV) or foliaceus (PF). Clinical features of the disease can mimic those seen in a drug reaction, erythema multiforme, Stevens-Johnson syndrome, pemphigus, lichen planus, or toxic epidermal necrolysis.

PNP/PAMS is associated in the majority of cases with non-Hodgkin lymphoma, chronic lymphocytic leukemia, thymoma, or Castleman disease. A serious complication includes bronchiolitis obliterans, which may lead to respiratory failure.

Reference Values

Report as positive or negative.

Negative in normal individuals.

Interpretation

In the appropriate clinical setting, a positive result can support a diagnosis of paraneoplastic pemphigus/paraneoplastic autoimmune multiorgan syndrome (PNP/PAMS). However, correlation with clinical features, histopathologic findings, results of serum studies (such as indirect immunofluorescence on monkey esophagus substrate and ELISA for Dsg1/3) is required for a final diagnosis. As the test is not entirely sensitive, a negative test result does not exclude the possibility of PNP/PAMS.

Cautions

Test results must be interpreted in the patient's individual clinical context.

Clinical Reference

1. Anhalt GJ, Kim SC, Stanley JR, et al: Paraneoplastic pemphigus. An autoimmune mucocutaneous disease associated with neoplasia. *N Engl J Med* 1990 Dec 20;323(25):1729-1735
2. Anhalt GJ, Aris-Abdo L, Bonitz P, Labib RS: Antigen specificity of paraneoplastic pemphigus: predictive value of diagnostic techniques based on the study of 17 patients and 135 control subjects. *J Invest Dermatol* 1992;98:580. Abstract
3. Liu AY, Valenzuela R, Helm TN, et al: Indirect immunofluorescence on rat bladder transitional epithelium: a test with high specificity for Paraneoplastic pemphigus. *J Am Acad Dermatol* 1993;28:696-699
4. Camisa C, Helm TN: Paraneoplastic pemphigus is a distinct neoplasia-induced autoimmune disease. *Arch Dermatol* 1993 Jul;129(7):883-886

Performance

Method Description

Commercially-prepared sections of rat bladder (substrate) are overlaid with patient serum; incubated, covered with fluorescein-conjugated IgG antiserum, and interpreted using fluorescent microscopy. (Unpublished Mayo method)

PDF Report

No

Day(s) Performed

Monday through Friday

Report Available

7 days

Specimen Retention Time

14 days

Performing Laboratory Location

Rochester

Fees and Codes**Fees**

- Authorized users can sign in to [Test Prices](#) for detailed fee information.
- Clients without access to Test Prices can contact [Customer Service](#) 24 hours a day, seven days a week.
- Prospective clients should contact their Regional Manager. For assistance, contact [Customer Service](#).

Test Classification

This test was developed and its performance characteristics determined by Mayo Clinic in a manner consistent with CLIA requirements. This test has not been cleared or approved by the U.S. Food and Drug Administration.

CPT Code Information

86255

LOINC® Information

| Test ID | Test Order Name | Order LOINC Value |
|---------|-------------------------------------|-------------------|
| PNPAB | Paraneoplastic Pemphigus, IgG Ab, S | 93233-5 |

| Result ID | Test Result Name | Result LOINC Value |
|-----------|-------------------------------------|--------------------|
| 61881 | Paraneoplastic Pemphigus, IgG Ab, S | 93233-5 |