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

<table>
<thead>
<tr>
<th>Gross hemolysis</th>
<th>Reject</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gross lipemia</td>
<td>Reject</td>
</tr>
<tr>
<td>Gross icterus</td>
<td>Reject</td>
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</table>

Specimen Stability Information

<table>
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<tr>
<th>Specimen Type</th>
<th>Temperature</th>
<th>Time</th>
<th>Special Container</th>
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<tbody>
<tr>
<td>Serum</td>
<td>Refrigerated (preferred)</td>
<td>14 days</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Frozen</td>
<td>30 days</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Ambient</td>
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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


Performance
Method Description
Commerically-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) and Time(s) Test Performed
Monday through Friday; 7 a.m.-5 p.m.

Analytic Time
7 days (Weekly testing)

Maximum Laboratory Time
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

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<th>Order LOINC Value</th>
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<tr>
<td>PNPAB</td>
<td>Paraneoplastic Pemphigus, IgG Ab, S</td>
<td>93233-5</td>
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<td>93233-5</td>
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