Overview

Useful For
Preferred screening test for patients suspected to have an autoimmune blistering disorder of the skin or mucous membranes (pemphigus)
Aiding in the diagnosis of pemphigus

Method Name
Enzyme-Linked Immunosorbent Assay (ELISA)

NY State Available
Yes

Specimen

Specimen Type
Serum

Specimen Required
Container/Tube:
Preferred: Red top
Acceptable: Serum gel

Specimen Volume: 1 mL

Specimen Minimum Volume
0.5 mL

Reject Due To

<table>
<thead>
<tr>
<th>Condition</th>
<th>Status</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gross hemolysis</td>
<td>OK</td>
</tr>
<tr>
<td>Gross lipemia</td>
<td>OK</td>
</tr>
<tr>
<td>Gross icterus</td>
<td>OK</td>
</tr>
</tbody>
</table>

Specimen Stability Information

<table>
<thead>
<tr>
<th>Specimen Type</th>
<th>Temperature</th>
<th>Time</th>
<th>Special Container</th>
</tr>
</thead>
<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>
<td>14 days</td>
<td></td>
</tr>
</tbody>
</table>

Clinical and Interpretive
**Clinical Information**

Pemphigus includes a group of often fatal autoimmune blistering diseases characterized by intraepithelial lesions. Pemphigus vulgaris and its variants may present with oral or mucosal lesions alone or with mucosal plus skin lesions. Pemphigus foliaceus and variants present with skin lesions alone.

Indirect immunofluorescence (IIF) studies reveal that both forms of pemphigus are caused by autoantibodies to cell surface antigens of stratified epithelia or mucous membranes and skin. These antibodies bind to calcium-dependent adhesion molecules in cell surface desmosomes, notably desmoglein 1 (DSG1) in pemphigus foliaceus and desmoglein 3 (DSG3) and/or DSG1 in pemphigus vulgaris. Desmogleins are protein substances located in and on the surface of keratinocytes. These proteins have been shown to be a critical factor in cell-to-cell adhesion. Antibodies to desmogleins can result in loss of cell adhesion, the primary cause of blister formation in pemphigus.

The diagnosis of pemphigus depends on biopsy and serum studies that characterize lesions and detect the autoantibodies that cause them. Originally, the serum studies were performed by IIF using monkey esophagus and other tissue substrates. The identification of the reactive antigens as DSG1 and DSG3 has made it possible to develop highly specific and sensitive enzyme-linked immunosorbent assay methods.

**Reference Values**

**DESMOGLEIN 1**

<20 RU/mL (negative)

> or =20 RU/mL (positive)

**DESMOGLEIN 3**

<20 RU/mL (negative)

> or =20 RU/mL (positive)

**Interpretation**

Antibodies to desmoglein 1 (DSG1) and desmoglein 3 (DSG3) have been shown to be present in patients with pemphigus. Many patients with pemphigus foliaceus, a superficial form of pemphigus have antibodies to DSG1. Patients with pemphigus vulgaris, a deeper form of pemphigus, have antibodies to DSG3 and sometimes DSG1 as well.

Antibody titer correlates in a semiquantitative manner with disease activity in many patients. Patients with severe disease can usually be expected to have high titers of antibodies to DSG. Titers are expected to decrease with clinical improvement.

Our experience demonstrates a very good correlation between DSG1 and DSG3 results and the presence of pemphigus. Adequate sensitivities and specificity for disease are documented. However, in those patients strongly suspected to have pemphigus either by clinical findings or by routine biopsy, and in whom the DSG assay is negative, the indirect immunofluorescence (CIFS / Cutaneous Immunofluorescence Antibodies [IgG], Serum) is recommended.

**Cautions**

Recommend repeat testing of indeterminate specimens, either with a fresh specimen drawn at a later time or the original specimen tested by another method.

The desmoglein 1 (DSG1) and desmoglein 3 (DSG3) results serve only as an aid to diagnosis and should not be
Test Definition: DSGAB
Desmoglein 1 and 3, Serum

interpreted as diagnostic by themselves. The results should be interpreted in conjunction with clinical evaluation of the patient along with other diagnostic procedures.

Performance of these assays in the pediatric population has not been established.

The assay performance characteristics have not been established for matrices other than serum.

A positive result indicates the presence of antibodies to recombinant DSG1 and DSG3 and does not specifically identify a certain type of pemphigus.

A negative result does not rule out the presence of pemphigus.

Clinical Reference

Performance

Method Description
This enzyme-linked immunosorbent assay (ELISA) method detects and measures serum levels of antibodies of certain pemphigus diseases. Calibrators and patient sera are added to microwells coated with desmoglein 1 (DSG1) and desmoglein 3 (DSG3) antigens, allowing antibodies to react with the immobilized antigens. After washing to remove any unbound serum proteins, horseradish peroxidase-conjugated IgG is added and incubated. Following another wash step, the peroxidase substrate is added and allowed to incubate for an additional period. Stop solution is then added to each well to cancel the enzyme reaction and to stabilize the color development. The assay can be quantified by measuring the reaction photometrically and plotting the results. The amount of antigen specific bound antibody is proportional to the color intensity. (Package inserts: Anti-Desmoglein 1 ELISA (IgG), Form EA_1495G_A_US_D03.doc, Version: 7/7/11; Anti-Desmoglein 3 ELISA (IgG), Form EA_1496G_A_US_D03.doc, Version: 7/7/11)

PDF Report
No

Day(s) and Time(s) Test Performed
Once or twice weekly, Monday through Friday; 7:30 a.m.-5 p.m., days of testing to be determined by the laboratory.
Test Definition: DSGAB
Desmoglein 1 and 3, Serum

Analytic Time
1 day

Maximum Laboratory Time
10 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 has been modified from the manufacturer's instructions. Its performance characteristics were 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
83516 x 2

LOINC® Information

<table>
<thead>
<tr>
<th>Test ID</th>
<th>Test Order Name</th>
<th>Order LOINC Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>DSGAB</td>
<td>Desmoglein 1 and 3, Serum</td>
<td>94335-7</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Result ID</th>
<th>Test Result Name</th>
<th>Result LOINC Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>606818</td>
<td>DSG 1</td>
<td>94336-5</td>
</tr>
<tr>
<td>606819</td>
<td>DSG 3</td>
<td>94337-3</td>
</tr>
</tbody>
</table>