Overview

Useful For
Confirming a diagnosis of pemphigoid, pemphigus, epidermolysis bullosa acquisita, or bullous lupus erythematosus

Method Name
Detection of IgG Anti-Intercellular Substance (ICS) and Anti-Basement Membrane Zone (BMZ) Antibodies by Indirect Immunofluorescence Technique Using Rhesus Monkey Esophagus Substrate and Human NaCl Split-Skin Substrate

NY State Available
Yes

Specimen

Specimen Type
Serum

Specimen Required

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

Specimen Volume: 2 mL

Reject Due To
- Gross hemolysis  OK
- Gross lipemia  Reject
- Gross icterus  OK

Specimen Minimum Volume
0.5 mL

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>
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</tbody>
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Clinical & Interpretive

Clinical Information
IgG anti-basement zone (BMZ) antibodies are produced by patients with pemphigoid. In most patients with bullous
pemphigoid, serum contains IgG anti-BMZ antibodies, while in cicatricial pemphigoid circulating IgG anti-BMZ antibodies are found in a minority of cases. Sensitivity of detection of anti-BMZ antibodies is increased when serum is tested using sodium chloride (NaCl)-split human skin as substrate.

Circulating IgG anti-BMZ antibodies are also detected in patients with epidermolysis bullosa acquisita (EBA) and bullous eruption of lupus erythematosus.

IgG anti-cell surface (CS) antibodies are produced by patients with pemphigus. The titer of anti-CS antibodies generally correlates with disease activity of pemphigus.

Reference Values
Report includes presence and titer of circulating antibodies. If serum contains BMZ antibodies on split-skin substrate, patterns will be reported as: 1) epidermal pattern, consistent with pemphigoid or 2) dermal pattern, consistent with epidermolysis bullosa acquisita.

Negative in normal individuals

Interpretation
Indirect immunofluorescence (IF) testing may be diagnostic when histologic or direct IF studies are only suggestive, nonspecific, or negative.

Anti-cell surface (CS) antibodies correlate with a diagnosis of pemphigus.

Anti-basement zone (BMZ) antibodies correlate with a diagnosis of bullous pemphigoid, cicatricial pemphigoid, epidermolysis bullosa acquisita (EBA), or bullous eruption of lupus erythematosus (LE).

If serum contains anti-BMZ antibodies, the pattern of fluorescence on sodium chloride(NaCl)-split skin substrate helps distinguish pemphigoid from EBA and bullous LE. Staining of the roof (epidermal side) or both epidermal and dermal sides of NaCl-split skin correlates with the diagnosis of pemphigoid, while fluorescence localized only to the dermal side of the split-skin substrate correlates with either EBA or bullous LE.

Cautions
Results should be interpreted in conjunction with clinical information, histologic pattern, and results of direct immunofluorescence (IF) study. In particular, the finding of low titer (< or =1:80) anti-CS antibodies should not be used alone (ie, without histologic or direct IF support) to confirm a diagnosis of pemphigus.

Clinical Reference

Performance

Method Description
Frozen sections of rhesus monkey esophagus and sodium chloride-split human skin are overlaid with dilutions of patient's serum, incubated, covered with fluorescein-conjugated IgG antiserum, and interpreted with a fluorescence microscope.

PDF Report
Test Definition: CIFS
Cutaneous Immfluor. Ab, S (IgG)

No

Specimen Retention Time
14 days

Performing Laboratory Location
Rochester

Fees & Codes

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 US Food and Drug Administration.

CPT Code Information
88346
88350