Autoimmune Blistering Diseases

ELISA Kits for the detection of Autoantibodies related to Pemphigus & Bullous Pemphigoid

Pemphigus
detection of anti-Desmoglein 1&3
MESACUP Desmoglein ELISA Kit
for Dsg1 & Dsg3

Bullous Pemphigoid
detection of anti-BP180 & BP230
MESACUP BP180 ELISA Kit
MESACUP BP230 ELISA Kit

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Autoimmune Blistering Diseases

Pemphigus and Bullous pemphigoid (BP) are distinct autoimmune blistering diseases that are characterized by the presence of autoantibodies directed against specific adhesion molecules of the skin and mucous membranes. Pemphigus is divided into two major subtypes: Pemphigus vulgaris (PV) and Pemphigus foliaceous (PF).

Pemphigus (PF, PV)
Pemphigus includes a group of often fatal, autoimmune, blistering diseases characterized by intraepithelial lesions. Pemphigus vulgaris and its variants may present with oral or other mucosal lesions alone or with mucosal plus skin lesions. Pemphigus foliaceous and its variants present with skin lesions alone.

Pemphigus Antigen Desmoglein
Indirect immunofluorescent (IIF) studies reveal that both forms of pemphigus are caused by autoantibodies to cell surface antigens of stratified epithelia of mucous membranes and skin. These antibodies bind to calcium dependent adhesion molecules in cell surface desmosomes, notably desmoglein 1 (Dsg1) in pemphigus foliaceous and desmoglein 3 (Dsg3) in pemphigus vulgaris. Pemphigus vulgaris patients with both mucosal and skin lesions have antibodies to both Dsg3 and Dsg1.

Bullous Pemphigoid (BP)
Bullous pemphigoid (BP) is an acquired autoimmune disease of the skin and mucous membranes, which is predominantly seen in the elderly. The skin lesions of these patients show separation of basal keratinocytes from the underlying dermis leading to subepidermal blisters.

Bullous Pemphigoid Antigen BP230 & BP180
Autoantibodies from patients with BP target two distinct molecules. One is a 230kDa molecule (BP230), which is localized ultrastructurally to the plaque of the hemidesmosome. The other is a 180kDa molecule (BP180), which is a type II-oriented transmembrane molecule with 15 collagenous and 16 non-collagenous extracellular domains, which has the C-terminal end outside of the cell. Because IgG cannot penetrate the cell membranes, BP180 is currently considered to be the target molecule to which pathogenic IgG autoantibodies bind. A number of studies have shown that a small noncollagenous region, called NC16A IgG domain, just outside of the membrane contain the major epitopes for BP180.

For Detection of Autoantibodies against Desmoglein 1 and 3

MESACUP Desmoglein TEST “Dsg1”
MESACUP Desmoglein TEST “Dsg3”

- High Specificity & Sensitivity
- Aids in diagnosis of pemphigus and distinction between PV and PF
- Parallel Fluctuation with Disease Activity
- Useful for monitoring disease activity in pemphigus & clinical transition between PV and PF

Anti-Dsg1 & Anti-Dsg3 titers in PF, PV and BP patients

<table>
<thead>
<tr>
<th>Test interpretation and expected value</th>
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</thead>
<tbody>
<tr>
<td>Anti-Dsg1 value (U/mL)</td>
</tr>
<tr>
<td>20 &amp; 14</td>
</tr>
<tr>
<td>20 &gt; 14</td>
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<tr>
<td>&gt; 14</td>
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</tbody>
</table>

| Anti-Dsg3 value (U/mL) | Interpretation |
| 20 | Positive |
| 20 > 7 | Indeterminate |
| > 7 | Negative |

PV: Pemphigus vulgaris, PF: Pemphigus foliaceus, BP: Bullous pemphigoid
**For Detection of Autoantibodies against BP180 & BP230**

**BP180 ELISA Kit**
**BP230 ELISA Kit**

![FDA cleared badge]

For In-Vitro Diagnostic Use Only

★ High Specificity > 98%
★ High Sensitivity

- Use NC16a as antigen: more sensitive and specific than immunoblotting or indirect immunofluorescence method

**Anti-BP180, BP230 IgG autoantibody titers by ELISA**

![Graph showing antibody titers]

**Sensitivity of BP180 & BP230 ELISA**

<table>
<thead>
<tr>
<th></th>
<th>BP Active (n=64)</th>
<th>BP Remission (n=175)</th>
<th>BP Total (n=239)</th>
</tr>
</thead>
<tbody>
<tr>
<td>BP180 Positive</td>
<td>84.40% (54/64)</td>
<td>64.60% (113/175)</td>
<td>69.90% (167/239)</td>
</tr>
<tr>
<td>BP230 Positive</td>
<td>64.10% (41/64)</td>
<td>78.90% (138/175)</td>
<td>74.90% (179/239)</td>
</tr>
<tr>
<td>BP180+BP230 Positive</td>
<td>95.30% (61/64)</td>
<td>98.90% (173/175)</td>
<td>97.90% (234/239)</td>
</tr>
</tbody>
</table>

P: Bullous pemphigoid
V: Pemphigus vulgaris
F: Pemphigus foliaceus
**Product Information**

**Brief assay procedure**

<table>
<thead>
<tr>
<th>Step</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Diluted sample (x101) 100 µL/well</td>
<td></td>
</tr>
<tr>
<td>1 hr (20-25°C)</td>
<td>Wash</td>
</tr>
<tr>
<td>1 hr (20-25°C)</td>
<td>Wash</td>
</tr>
<tr>
<td>30 min (20-25°C)</td>
<td>Substrate 100 µL/well</td>
</tr>
<tr>
<td>Read absorbance 450nm</td>
<td>Stop solution 100 µL/well</td>
</tr>
</tbody>
</table>

- Recombinant antigens
- Anti-Dsg1 or Anti-Dsg3 or Anti-BP180
- HRP-conjugated anti-human IgG

**Kit components**

- Microwell Strips coated with antigens
- Calibrator 1 (0 U/mL)*
- Calibrator 2 (100 U/mL)*
- Conjugate Reagent (HRP-conjugated anti-human IgG)
- Conjugate Diluent**
- Assay Diluent*
- Wash Concentrate (10x)
- Substrate*
- Stop Solution*

* Ready for use
** Provided in MESACUP Desmoglein TEST (Dsg1, Dsg3)

**Breakable Assay Strip!**

*Easy to operate & Economical use*

**References**