### Epithelial Antibody-Associated Immunobullous Diseases Testing

#### Indications for Testing

**Symptomatic Adults**
- See additional indications for testing for symptomatic adults with neoplastic disease and symptomatic pregnant women (typically in 2nd or 3rd trimester).

**Test for Immunobullous Diseases** *(pemphigus, pemphigoid, EBA, linear IgA disease, or dermatitis herpetiformis)*

- **Obtain biopsy specimen(s) of intact skin or mucosa**
  - Lesional for histopathology
  - Perilesional for cutaneous Direct Immunofluorescence, Biopsy

- **Obtain serum specimen for Immunobullous Disease Panel, Epithelial**
  - OR, if strong clinical suspicion for pemphigoid, order Basement Membrane Zone Antibody Panel (BMZ) and IgG antibodies, IgG BMZ antibodies, IgA BMZ antibodies, Epidermal and Dermal, or combined Epidermal/Dermal pattern antibodies
  - OR, if strong clinical suspicion for dermatis herpetiformis, order Basement Membrane Zone Antibody Panel,

**Repeat testing:**
- In approximately 3-6 mos for persistent unexplained disease
- In approximately 6-12 wks for rapidly evolving or changing disease

**Monitor for developing immunobullous disease**

- Consider porphyria or pseudoporphyria (perilesional skin biopsy is not specific but will not be consistent with immunobullous disease)

**Consider treatment status and test repetition, as dietary gluten restriction may reduce/eliminate detectable antibodies, and/or consider seronegative disease**

**Negative**

**Positive**

**Consider dermatitis herpetiformis/celiac disease serologies**

**Negative**

**Positive**

**Testing will likely demonstrate antibodies to bullous pemphigoid antigens (180 kDa and 230 kDa), and/or IgG (BP 180, BP 230)**

#### Abbreviations
- BMZ: Basement membrane zone
- DIF: Direct immunofluorescence
- EBA: Epidermolysis bullosa acquisita
- ELISA: Enzyme-linked immunosorbent assay
- eTG: Epidermal transfugaminase
- IFA: Indirect immunofluorescence assay
- Ig: Immunoglobulin
- tTG: Tissue transglutaminase

#### Order

**Pemphigus**

**IgA pemphigus**

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*The Basement Membrane Zone Antibody Panel includes epithelial IgG and IgA BMZ antibody testing by IFA as well as bullous pemphigoid antigens, BP 180 and BP 230, IgG antibody and type VII collagen IgG antibody testing by ELISA. These same test components are included in the Immunobullous Disease Panel, Epithelial, along with cell surface (pemphigus) antibody testing components. The Pemphigoid Antibody Panel – Epithelial Basement Membrane Zone Antibodies, IgG and IgA, BP 180 and BP 230 Antibodies, IgG, can be ordered separately (but this panel does not include type VII collagen IgG antibody testing by ELISA), and the IFA and ELISA components can be ordered separately. IFA testing is determined on BMZ split skin substrate, also known as salt-split skin.*

*The Celiac Disease Reflexive Cascade includes total IgA; celiac disease dual antigen screen; deamidated gliadin peptide antibody, IgA and IgG; Tissue TG, IgA and IgG by ELISA; and endomysial antibody, IgA by IFA.*

*Antigens used in this assay include desmoglein 1 (320 kDa), desmoglein 3 (270 kDa), and type VII collagen (200 kDa).*