Screen for Immunobullous Diseases (pemphigus, pemphigoid, epidermolysis bullosa acquista, linear IgA disease, or dermatitis herpetiformis)
- Histology
- Perilesional skin biopsy
- Cutaneous Direct Immunofluorescence, Biopsy
- Serology
  - Pemphigus Antibody Panel - Epithelial Cell Surface Antibodies and Desmoglein 1 and Desmoglein 3 Antibodies, IgG
  - Pemphigoid Antibody Panel - Epithelial Basement Membrane Zone Antibodies, IgG and IgA, BP180 and BP230 Antibodies, IgG
  - Tissue Transglutaminase (tTG) Antibody, IgA with Reflex to Endomysial Antibody, IgA by IFA (alternative test: Celiac Disease Dual Antigen Screen with Reflex)
  - Epithelial Skin Antibody AND Tissue Transglutaminase (tTG) Antibody, IgA with Reflex to Endomysial Antibody, IgA by IFA

**Screen for Immunobullous Diseases**
- Repeat screening
- Consider porphyria or pseudoporphyria (perilesional skin biopsy is not specific but will not be consistent with immunobullous disease)
- Repeat screening:
  - In 3-6 months for persistent unexplained disease
  - In 6-12 weeks for rapidly evolving disease

**Cutaneous DIF skin biopsy**
- Negative
- Positive
  - IgA endomysial antibodies*
  - IgA basement membrane zone epidermal, dermal or combined epidermal/dermal pattern antibodies
- Monitor for developing immunobullous disease
  - Consider porphyria or pseudoporphyria (perilesional skin biopsy is not specific but will not be consistent with immunobullous disease)
- Repeat screening:
  - In 3-6 months for persistent unexplained disease
  - In 6-12 weeks for rapidly evolving disease

**Cutaneous DIF skin biopsy**
- Negative
- Positive
  - Monitor treatment response with Epithelial Basement Membrane Zone Antibody IgA
  - Epithelial Skin Antibody

**Linear IgA disease**
- Monitor treatment response with IgA endomysial*, Tissue Transglutaminase (tTG) Antibody, IgA; and Epidermal Transglutaminase (etG/tTG) Antibody, IgA by ELISA
  - (or if IgA deficient Tissue Transglutaminase Antibody, IgG)

**Indications for Testing**
- Symptomatic adults

**Presence of neoplastic disease**
- ORDER
  - Perilesional skin biopsy for Cutaneous Direct Immunofluorescence, Biopsy
  - Paraneoplastic Pemphigus Antibody Screen

**Immunobullous Skin Diseases Testing**
- Negative
- Positive, titer >1:10

**Pemphigus**
- IgA endomysial antibodies*
- IgA basement membrane zone epidermal, dermal or combined epidermal/dermal pattern antibodies

**Linear IgA disease**
- Monitor treatment response with Epithelial Basement Membrane Zone Antibody IgA
  - Epithelial Skin Antibody
- Dermatitis herpetiformis

**Pemphigoid**
- Epidermolysis bullosa acquista

**ORDER**
- Collagen Type VII Antibody IgG by ELISA

**ORDER**
- Bullous Pemphigoid Antigens (180 kDa and 230 kDa), IgG (BP180, BP230 antibodies) (Note: Necessary if pemphigoid panel testing was not ordered)

**ORDER**
- Monitor treatment response with Epithelial Basement Membrane Zone Antibody IgG
  - Epithelial Skin Antibody
  - Collagen Type VII Antibody IgG by ELISA

**ORDER**
- Monitor treatment response with Pemphigoid Antibody Panel - Epithelial Basement Membrane Zone Antibodies, IgG and IgA, BP180 and BP230 Antibodies, IgG
- Monitor treatment response with Pemphigoid Antibody Panel - Epithelial Cell Surface Antibodies and Desmoglein 1 and Desmoglein 3 Antibodies, IgG

**Positive**
- P. foliaceus (most common)
- P. vulgaris (most common)
- P. erythematous
- P. vegetans

**Pregnant female, typically 2nd or 3rd trimester**
- ORDER
  - Perilesional skin biopsy for Cutaneous Direct Immunofluorescence, Biopsy
  - Herpes Gestationis Factor (Complement-Fixing Basement Membrane Zone Antibody IgG) and IgG BP180 antibody level

**Negative**
- Monitor treatment response with Herpes Gestationis Factor (Complement-Fixing Basement Membrane Zone Antibody IgG) and IgG BP180 antibody level

* For Endomysial Antibodies, a screening IgA tissue transglutaminase assay (ELISA) can be ordered OR request Endomysial Antibody test by indirect immunofluorescence (IgA and/or IgG) with or without accompanying IgA and/or IgG tissue transglutaminase assay through the Immunodermatology Laboratory.

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