

Pemphigus Panel

IGG EPITHELIAL CELL SURFACE ANTIBODIES AND LEVELS OF IGG DESMOGLEIN 1 AND DESMOGLEIN 3 ANTIBODIES IN SERUM

DIAGNOSTIC TESTS FOR PEMPHIGUS, AN IMMUNOBULLOUS DISEASE OF SKIN AND MUCOUS MEMBRANES

Clinical Background

- Pemphigus is a group of autoimmune blistering diseases affecting skin and mucous membranes. Prior to the introduction of glucocorticoids and other immunosuppressive therapies, it was commonly fatal.
- In pemphigus, IgG is deposited on the cell surfaces (formerly referred to as intercellular substance) of epidermal and stratified squamous epithelial cells in and around affected areas.
- IgG cell surface antibodies circulate in the blood in up to 80 percent of patients with pemphigus.
- Desmoglein antibodies in serum from individuals with pemphigus bind to calcium-dependent adhesion molecules in cell surface desmosomes of epithelium.
- IgG desmoglein 1 autoantibodies are present in patients with pemphigus foliaceus (and likely pemphigus erythematosus), and IgG desmoglein 3 autoantibodies are present in patients with pemphigus vulgaris (and likely pemphigus vegetans).
- Desmoglein antibody levels in serum correlate with disease activity. In recent years, these antibodies have been shown to be pathogenic and are not simply epiphenomena of the diseases.
- Diagnosis of immune-mediated diseases of skin and other epithelial organs is aided by specific immunodermatologic testing. Although the various cutaneous immunobullous diseases have certain characteristic features that distinguish them, often the clinical presentation is atypical and shows overlap with other diseases.
- Testing for IgG cell surface and IgG desmoglein antibodies is important in establishing the diagnosis of pemphigus; this testing is highly sensitive and specific. In addition, IgG desmoglein antibody testing can distinguish pemphigus foliaceus from pemphigus vulgaris and from other immune-mediated skin disease. Moreover, because antibody titers correlate with disease activity, the results can be used to monitor disease expression and response to treatment.

Methodology

- IgG cell surface antibodies to human epidermis and monkey esophagus epithelium are detected by indirect immunofluorescence (IFA). Serial twofold dilutions of serum in calcium buffer, incubated with substrates and followed by fluoresceinated antibody to human IgG, are used to screen for the presence of antibodies. When positive, limiting dilutions are carried out for endpoint titers to establish levels of circulating antibodies. (IgA cell surface antibodies can also be tested by ordering test number 0092106.)
- Enzyme linked immunosorbent assays (ELISA) are used to detect IgG desmoglein 1 and IgG desmoglein 3 antibodies in a patient's serum.

Indications for Ordering

- Testing for IgG cell surface and desmogleins 1 and 3 antibodies is appropriate for patients suspected of having or known to have any type of pemphigus.
- Cell surface antibody testing by IFA is a well-established test and is also available to detect IgA pemphigus antibodies (0092106). Detection of relative levels of desmoglein antibodies aids in the diagnosis and subtyping of pemphigus, distinguishing pemphigus from other immune-mediated disease, and in monitoring disease activity.
- Because low titer cell surface antibodies may rarely be found in normal individuals and in patients with burns, drug interactions, and fungal infections, interpretation of results should be done along with clinical correlation.

Interpretation

- IgG cell surface serum antibodies present in titers > 1:10 are positive.
- Desmoglein 1 and desmoglein 3 IgG antibodies in serum are interpreted as follows:
 - **IgG desmoglein 1 antibody level:**
 - Negative: Fewer than 14 units
 - Borderline/indeterminate: 14–20 units
 - Positive: Greater than 20 units
 - **IgG desmoglein 3 antibody level:**
 - Negative: Fewer than 9 units
 - Borderline/indeterminate: 9–20 units
 - Positive: Greater than 20 units
 - IgG desmoglein 1 antibodies predominate in pemphigus foliaceus.
 - IgG desmoglein 3 antibodies predominate in pemphigus vulgaris.
- Cell surface antibodies are negative in normal individuals. Titers correlate with disease activity. IgG desmoglein antibodies are negative in normal individuals. Although the predominate antibodies differentiate the subtypes of pemphigus, some overlap may occur. Patients with both skin and mucosal lesions may have IgG antibodies to both desmogleins 1 and 3. Desmoglein antibody levels correlate with disease activity and may be low or negative in treated disease. Patients with results in the borderline/indeterminate range should be monitored for disease activity. Desmoglein antibodies may or may not be positive in paraneoplastic pemphigus; however, indirect immunofluorescence is often positive for cell surface antibodies in paraneoplastic pemphigus.

Related Test

[Pemphigus IgA Antibodies \(0092106\)](#)

References

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2. Mutasim DF, Adams BB. Immunofluorescence in dermatology. *J Am Acad Dermatol* 2001;45:803–22.
3. Plager DA, et al. Structural and functional cutaneous immunology. In *Middleton's Allergy: Principles and Practice*, 6th ed. Adkinson NF, et al, eds. 2003; Philadelphia: Mosby Inc. 671–86.
4. Harman KE. New laboratory techniques for the assessment of acquired immunobullous disorders. *Clin Exp Dermatol* 2002;27:40–6.
5. Anhalt GJ, Diaz LA. Research advances in pemphigus. *JAMA* 2001;285:652–4.

Test Information

0090650

Pemphigus Panel IgG Epithelial Cell Surface Antibodies and Levels of IgG Desmoglein 1 and Desmoglein 3 Antibodies, Serum

For specific collection, transport, and testing information, refer to the ARUP Web site at www.aruplab.com.

For information on test selection, ordering, and interpretation, refer to ARUP Consult® at www.arupconsult.com.