

Desmoglein 1 and Desmoglein 3 IgG Antibodies in Serum

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

Introduction

- 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 and are detected with immunoassays.
- IgG 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). IgG desmoglein 3 autoantibodies are present in patients with pemphigus vulgaris (and likely pemphigus vegetens). IgG 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.
- 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 not typical and shows overlap with other diseases.
- Testing for IgG desmoglein antibodies is an important aid in establishing the diagnosis of pemphigus and is highly sensitive and specific. In addition, desmoglein antibody testing can distinguish pemphigus foliaceus from pemphigus vulgaris and other immune-mediated skin disease. Moreover, because antibody titers correlate with disease activity, the tests can be used to monitor disease activity and response to treatment.

Indications for Ordering

- Testing for IgG desmoglein 1 and IgG desmoglein 3 antibody levels is appropriate for patients suspected of having or known to have any type of pemphigus.
- This test may not be positive in paraneoplastic pemphigus but may help distinguish it from other types of pemphigus.
- Detection of relative levels of desmoglein antibodies aids in the diagnosis of and subtyping of pemphigus, distinguishing pemphigus from other immune-mediated disease, and monitoring disease activity.

Laboratory Diagnosis

- 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.
- 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. This test may or may not be positive in paraneoplastic pemphigus.

Methodology

Enzyme linked immunosorbent assay (ELISA) is used to detect and measure IgG desmoglein 1 and IgG desmoglein 3 antibody levels in patient's serum.

References

1. Harman KE. New laboratory techniques for the assessment of acquired immunobullous disorders. *Clin Exp Dermatol* 2002;27:40–6.
2. Anhalt GJ, Diaz LA. Research advances in pemphigus. *JAMA* 2001;285:652–4.
3. Amagai M, et al. Usefulness of enzyme linked immunosorbent assay using recombinant desmogleins 1 and 3 for serodiagnosis of pemphigus. *Brit J Dermatol* 1999;140:351–7.
4. Amagai M, et al. The clinical phenotype of pemphigus is defined by the anti-desmoglein autoantibody profile. *J Am Acad Dermatol* 1999; 40:167–70.

Test Information

0090649 **IgG Desmoglein 1 & Desmoglein 3**

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.