MAYO CLINIC LABORATORIES

Test Definition: DSGAB

Desmoglein 1 (DSG1) and Desmoglein 3 (DSG3), IgG Antibodies, Serum

Overview

Useful For

Preferred screening test for patients suspected of having an autoimmune blistering disorder of the skin or mucous membranes (pemphigus)

Aiding in the diagnosis of pemphigus

Testing Algorithm

For information see Autoimmune Blistering Disease Diagnostic Algorithm.

Special Instructions

- Autoimmune Blistering Disease Diagnostic Algorithm

Method Name

Enzyme-Linked Immunosorbent Assay (ELISA)

NY State Available

Yes

Specimen

Specimen Type

Serum

Specimen Required

Supplies: Sarstedt Aliquot Tube, 5 mL (T914)
Collection Container/Tube:
Preferred: Red top
Acceptable: Serum gel
Submission Container/Tube: Plastic vial
Specimen Volume: 1 mL Serum
Collection Instructions: Centrifuge and aliquot serum into a plastic vial.

Specimen Minimum Volume

Serum: 0.5 mL

Reject Due To

Gross	OK

hemolysis	
Gross lipemia	OK
Gross icterus	OK

Specimen Stability Information

Specimen Type	Temperature	Time	Special Container
Serum	Refrigerated (preferred)	14 days	
	Ambient	14 days	
	Frozen	30 days	

Clinical & Interpretive

Clinical Information

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

Indirect immunofluorescence studies reveal that both forms of pemphigus are caused by autoantibodies to cell surface antigens of stratified epithelia or mucous membranes and skin. These antibodies bind to calcium-dependent adhesion molecules in cell surface desmosomes, notably desmoglein 1 (DSG1) in pemphigus foliaceus and desmoglein 3 (DSG3) or DSG1 in pemphigus vulgaris. Desmogleins are protein substances located in and on the surface of keratinocytes. These proteins have been shown to be a critical factor in cell-to-cell adhesion. Antibodies to desmogleins can result in loss of cell adhesion, the primary cause of blister formation in pemphigus.

The diagnosis of pemphigus depends on biopsy and serum studies that characterize lesions and detect the autoantibodies that cause them. Originally, the serum studies were performed by IIF using primate esophagus and other tissue substrates. The identification of the reactive antigens as DSG1 and DSG3 has made it possible to develop highly specific and sensitive enzyme-linked immunosorbent assay methods.

Reference Values

Desmoglein 1:
<20 RU/mL (Negative)
> or =20 RU/mL (Positive)

Desmoglein 3:
<20 RU/mL (Negative)
> or =20 RU/mL (Positive)

Interpretation

Antibodies to desmoglein 1 (DSG1) and desmoglein 3 (DSG3) have been shown to be present in patients with pemphigus. Many patients with pemphigus foliaceus, a superficial form of pemphigus have antibodies to DSG1. Patients with pemphigus vulgaris, a deeper form of pemphigus, have antibodies to DSG3 and sometimes DSG1 as well.

Antibody titer correlates in a semiquantitative manner with disease activity in many patients. Patients with severe disease can usually be expected to have high titers of antibodies to DSG. Titers are expected to decrease with clinical improvement.

Our experience demonstrates a very good correlation between DSG1 and DSG3 results and the presence of pemphigus. Adequate sensitivities and specificity for disease are documented (see Supportive Data). However, in those patients strongly suspected to have pemphigus either by clinical findings or by routine biopsy, and in whom the DSG assay is negative, indirect immunofluorescence testing is recommended. For more information see CIFS / Cutaneous Immunofluorescence Antibodies, IgG and IgG4, Serum.

Cautions

Recommend repeat testing of indeterminate specimens, either with a fresh specimen collected at a later time or the original specimen tested by another method.

The desmoglein 1 (DSG1) and desmoglein 3 (DSG3) results serve only as an aid to diagnosis and should not be interpreted as diagnostic by themselves. The results should be interpreted in conjunction with clinical evaluation of the patient along with other diagnostic procedures.

Performance of these assays in the pediatric population has not been established.

The assay performance characteristics have not been established for matrices other than serum.

A positive result indicates the presence of antibodies to recombinant DSG1 and DSG3 and does not specifically identify a certain type of pemphigus.

A negative result does not rule out the presence of pemphigus.

Clinical Reference

1. Amagai M, Tsunoda K, Zillikens D, Nagai T, Nishikawa T. The clinical phenotype of pemphigus is defined by the anti-desmoglein autoantibody profile. J Am Acad Dermatol. 1999;40(2 Pt 1):167-170
2. Amagai M, Komai A, Hashimoto T, et al. Usefulness of enzyme-linked immunoabsorbent assay using recombinant desmogleins 1 and 3 for sero-diagnosis of pemphigus. Brit J Dermatol. 1999;140(2):351-357
3. Harman KE, Gratian MJ, Bhogal BS, Challacombe SJ, Black M. The clinical significance of autoantibodies to desmoglein 1 in 78 cases of pemphigus vulgaris. J Invest Derm. 1999;112(4):568. Abstract 273
4. Harman KE, Gratian MJ, Seed PT, Bhogal BS, Challacombe SJ, Black MM. Diagnosis of pemphigus by ELISA: a critical evaluation of two ELISAs for the detection of antibodies to the major pemphigus antigens, desmoglein 1 and 3. Clin Exp Dermatol. 2000;25(3):236-240
5. Prussmann W, Prussmann J, Koga H, et al. Prevalence of pemphigus and pemphigoid autoantibodies in the general population. Orphanet J Rare Dis. 2015;10:63
6. Toosi S, Collins JW, Lohse CM, et al. Clinicopathologic features of IgG/IgA pemphigus in comparison with classic (IgG) and IgA pemphigus. Int J Dermatol. 2016;55(4):e184-e190
7. Montagnon CM, Tolkachjov SN, Murrell DF, Camilleri MJ, Lehman JS. Intraepithelial autoimmune blistering dermatoses: Clinical features and diagnosis. J Am Acad Dermatol. 2021;84(6):1507-1519

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Performance

Method Description

This enzyme-linked immunosorbent assay detects and measures serum levels of antibodies of certain pemphigus diseases. Calibrators and patient sera are added to microwells coated with desmoglein 1 and desmoglein 3 antigens, allowing antibodies to react with the immobilized antigens. After washing to remove any unbound serum proteins, horseradish peroxidase-conjugated IgG is added and incubated. Following another wash step, the peroxidase substrate is added and allowed to incubate for an additional period. Stop solution is then added to each well to cancel the enzyme reaction and to stabilize the color development. Quantification is performed by measuring the reaction photometrically and plotting the results. The amount of antigen specific bound antibody is proportional to the color intensity.(Package inserts: Anti-Desmoglein 1 ELISA [IgG], Form EA_1495G_A_US_D04. EuroImmun; 07/08/2020; Anti-Desmoglein 3 ELISA [IgG], Form EA_1496G_A_US_D04. EuroImmun; 07/08/2020)

PDF Report

No

Day(s) Performed

Varies

Report Available

1 to 10 days

Specimen Retention Time

30 days

Performing Laboratory Location

Mayo Clinic Laboratories - Rochester Main Campus

Fees & Codes

Fees

- Authorized users can sign in to Test Prices for detailed fee information.
- Clients without access to Test Prices can contact Customer Service 24 hours a day, seven days a week.
- Prospective clients should contact their account representative. For assistance, contact Customer Service.

Test Classification

This test has been modified from the manufacturer's instructions. Its performance characteristics were determined by Mayo Clinic in a manner consistent with CLIA requirements. This test has not been cleared or approved by the US Food and Drug Administration.

CPT Code Information

83516 x 2

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LOINC® Information

Test ID	Test Order Name	Order LOINC® Value
DSGAB	Desmoglein 1 and 3, Serum	94335-7

Result ID	Test Result Name	Result LOINC® Value
606818	DSG 1	94336-5
606819	DSG 3	94337-3