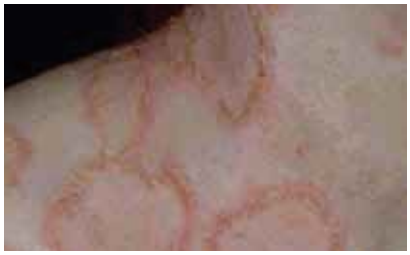
immco DIAGNOSTICS
The total solution in autoimmunity.™

Pemphigus Vulgaris

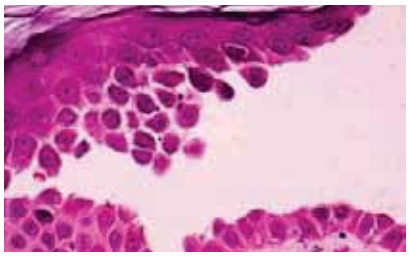
Pemphigus refers to a group of potentially life-threatening autoimmune blistering diseases of the skin and mucous membranes. Three primary subsets of pemphigus have been identified and include pemphigus vulgaris (PV), pemphigus foliaceus, and paraneoplastic pemphigus. Each type of pemphigus has distinct clinical and immunopathologic features. Pemphigus vulgaris (PV) accounts for approximately 70% of pemphigus cases. Male-to-female ratio is approximately equal and the mean age of onset is approximately 50-60 years.

Clinical Features

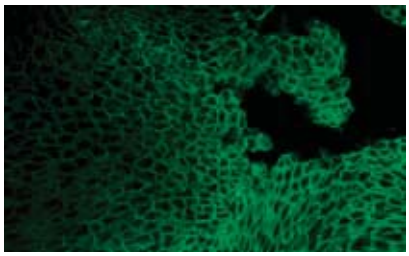
- The pathophysiology of PV is characterized by circulating autoantibodies directed against intercellular antigens Desmoglein 1 and Desmoglein 3 (DSg1 and DSg3)
- The binding of autoantibodies results in a loss of cell-cell adhesion, a process termed acantholysis
- Patients have ill-defined, irregularly shaped, gingival, buccal or palatine erosions, which are painful and slow to heal
- Mucosal lesions may precede cutaneous lesions by months



Blisters on the skin



Acantholysis and supra-epithelial separation



Intercellular immunodeposits of IgG in the squamous epithelial cells

Diagnosis

To establish a diagnosis of PV, histopathology studies are done on a biopsy taken from the edge of a blister:

- Histopathology can demonstrate an intradermal blister
- Direct immunofluorescence (IF) studies on normal-appearing (perilesional) skin or mucosa and indirect IF serum studies are required
- Direct IF demonstrates IgG intercellular deposition throughout the epidermis
- Circulating intercellular antibodies to DSg1 and DSg3 are detected in almost all of patients with PV depending on the extent of the lesions
- The titer of circulating antibody may correlate with disease course

Why choose Immco Diagnostics?

- Immco was co-founded by periodontists
- Over 40 years experience providing specialized Oral and Maxillofacial pathology biopsy services
- Board certified oral and immuno pathologists
- Accuracy and reliability
- Consultations and second opinions
- More than 100 published articles on autoimmune mediated periodontal diseases
- Lab report within 48 hours of sample receipt via fax, mail or **Immco Online**, a HIPAA-compliant web tool

Incidence of Intercellular Antibodies in Pemphigus

Substrate	No. Tested	No. Positive	% Positive
Monkey Esophagus	123	111	89
Guinea Pig Esophagus	123	103	81
Monkey / Guinea Pig Esophagus	123	122	99

Optimal Biopsy Site for Immunological Investigations

Tissue	Site
Skin or Mucosa	Take one biopsy from a perilesional area, adjacent to active or new blister
	Take one biopsy from an adjacent or normal area at least 3 mm from a lesion

Treatment

Current treatment is largely based on systemic immunosuppression using corticosteroids, with azathioprine or other adjuvants. Newer therapies with potentially fewer adverse effects appear promising.

Immco Tests

Code Description

510 Direct Immunofluorescence – Routine panel tests for the presence of IgG, IgA, IgM, Fibrin, C3 plus C5b-9 and/or IgG4, depending on diagnosis.
Methodology: DIF (Direct Immunofluorescence)
Reference Range: Detailed interpretation accompanies report.
CPT Code: 88346(x6 or x7)
Turnaround Time: Report availability is within 48 hours from the time of specimen receipt.

Sample Submission

Specimen collection kits are available free of charge. Please call 1.800.537.8378 or e-mail laboratoryservices@immco.com for supplies.

Use appropriate tube(s) as follows:

Immunofluorescence:
Lesional biopsy.....Red tube
Normal biopsy.....Purple tube
H&E biopsy .....Green tube
Serology.....Orange tube

Specimen can be shipped by courier services, U.S. Postal service and overnight carriers free of charge. Results are reported within two business days of the receipt of the specimen via mail, fax and through Immco online, a HIPAA-compliant web tool at www.immco.com.

Selected References

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Suresh L, Neiders ME. Definitive and differential diagnosis of desquamative gingivitis through direct immunofluorescence studies. J Periodontol. 2012; 83:1270-8.

Tron F, Gilbert D, Mouquet H, Joly P, Drouot L, Makni S, Masmoudi H, Charron D, Zitouni M, Loiseau P, Ben Ayed M. Genetic factors in pemphigus. J Autoimmun. 2005; 24:319-28.

For details about our products and services, please contact laboratoryservices@immco.com.

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United States
Immco Diagnostics, Inc.
10 Earhart Drive
Suite 100
Williamsville, NY 14221

+1.716.691.0091
800.537.8378
+1.716.691.6955 Fax
laboratoryservices@immco.com
www.immco.com

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