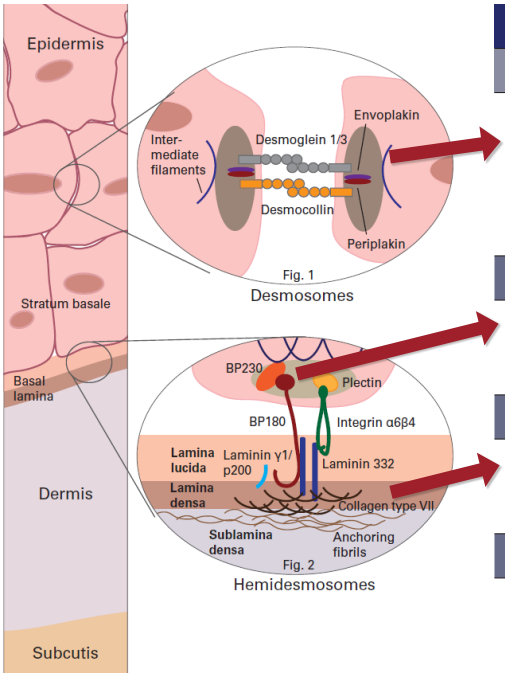
THE PATHCARE NEWS

SEROLOGICAL TESTING FOR AUTOIMMUNE BULLOUS DERMATOSES

Autoimmune bullous skin disease (AIBD) is a group of blistering skin disorders caused by autoantibodies directed against various structural proteins of the skin. The main clinical features are blisters and bullae in the skin and mucous membranes, progressing to erosive lesions after rupture. Based on the pathogenesis, bullous skin diseases can be divided into autoimmune and non-autoimmune forms. Pathogenic autoantibodies can be detected in both the circulation and skin lesions in autoimmune bullous diseases.



Classification of Autoimmune Bullous Dermatoses

Pemphigus diseases

Intraepidermal blistering due to antibodies directed against *desmosomes* (**Desmoglein 1/3**) that cause disruptions of intercellular connections in the prickle-cell layer of the epidermis of the outer skin and mucous membranes.

Pemphigus conditions include *pemphigus vulgaris (PV), pemphigus foliaceous (PF), paraneoplastic pemphigus (PNP), and IgA pemphigus.*

Pemphigoid diseases

Subepidermal blistering due to antibodies directed against *hemidesmosome* components (**BP230 and BP180**) and structural filaments. Pemphigoid conditions include *bullous pemphigoid (BP), gestational pemphigoid (GP), mucosal pemphigoid, and linear IgA dermatosis.*

Epidermolysis bullosa acquisita (EBA)

A severe autoimmune blistering dermatosis with autoantibodies in the inflammatory form targeting **collagen type VII**, which presents with **subepidermal blister** formation in the outer skin and mucous membranes.

Dermatitis herpetiformis (DH)

Considered as the cutaneous manifestation of coeliac disease with **subepidermal blistering** and unaffected mucous membranes. It is marked by autoantibodies against endomysium (EMA) and tissue transglutaminase (tTG). Symptoms typically improve with a gluten-free diet.

Figure 1. Schematic representation of autoimmune bullous dermatoses autoantibody targets. Adapted from Saschenbrecker S *et al.*[1]

Laboratory confirmation of the diagnosis

1. Histopathology – on skin or mucosa biopsy.
2. Direct immunofluorescence (DIF) - microscopy of perilesional skin tissue.
3. Serological tests - detection of autoantibodies via indirect immunofluorescence assay (IFA).

Histology and DIF are essential, but biopsies are invasive, and representative sampling may be challenging. Serological tests are less invasive and provide valuable supplementary information for diagnosing and classifying AIBD.

Serological screening using tissue substrates

Indirect immunofluorescence assays (IFA) employ primate oesophagus to identify autoantibodies against desmosomes (intraepidermal conditions) or hemidesmosomes (subepidermal conditions). Additionally, salt-split skin is used to differentiate various subepidermal blistering diseases by determining the location of the antibody binding – either on the epidermal side (e.g., bullous pemphigoid), or dermal side (e.g., epidermolysis bullosa acquisita). Recombinant monospecific substrates, using either transfected cells or recombinant antigens coated directly onto BIOCHIPs, are included in the test profile for the qualitative detection of Desmoglein 1 and 3 (Dsg1/Dsg3) as well as BP180 and BP230.

Serological testing for AIBD is available at Pathcare and includes Desmoglein 1 and 3 (Dsg1/Dsg3) and BP180 and BP230.
Collagen Type VII IgG antibody testing can be arranged on special request.

Request test:	**P5781** ☑ Autoimmune bullous dermatosis serology	
Sample type:	**SST** (Serum separator/yellow top tube)	**TAT**: 7 days from reaching the reference laboratory
Cost:	Please enquire at your local laboratory. Cost may differ depending on payment method.	

Ref: Saschenberger S et al. Serological diagnosis of autoimmune bullous skin diseases. Frontiers Immunol 2019; 10(1974).
Prepared by: J van Wyk and M Lloyd