**SOUHRN DISERTAČNÍ PRÁCE V ANGLICKÉM JAZYCE**

Autoimmune bullous diseases are severe and chronic conditions, which involve skin and mucosal surface. The etiology is unknown. The specific antibodies against structural components of cellular adhesions molecules of epidermis, at the dermal-epidermal junctions or at the basement membrane zone, are characteristic. The connection between antibodies and targeted antigens leads to cell-cell or cell-matrix discontinuity, which develops into blister formation. Intraepidermal disruption is typical for pemphigus diseases, dermo-epidermal (sub-epidermal) blistering process is specific for pemphigoid diseases. Detection of specific autoantibodies either tissue-bound or circulating in serum is essential to diagnose autoimmune nature of autoimmune bullous disease.

The specific antibodies for bullous pemphigoid against the hemidesmosomal antigens BP 180, BP 230 are seen, which connect the basal keratinocytes to basement membrane. The correct diagnosis is based on positivity of minimally 2 markers - histological proof, detecting of tissue-bound antibody Immunoglobulin G and C3 component of complement at the basement membrane zone by direct immunofluorescence, detection of circulating Immonoglobulin G by indirect imunofluorescence on the monkey or rabbit esophagus or salt-split skin, as well as by BP180, BP230 ELISA. Typical clinical picture can help to make the right diagnosis.

Other diagnostic method, which was commonly used just in the diagnostics of pemphigoid gestationis (originally named herpes gestationis) in the 1970s is complement fixation test (herpes gestationis factor test). It has not been published yet for the greater group of bullous pemphigoid patients. Complement fixation test was assessed in 300 bullous pemphigoid positive patients compared to 136 negative controls.

Indirect immunofluorescence is one of the diagnostic methods with relatively low sensitivity itself. Among bullous pemphigoid patients is already well-known, that all Immunoglobulin G subclasses play a role (Immunoglobulin G1-3 as complement fixing antibodies, Immunoglobulin G4 can not fix complement). We deeply targeted each subtypes in 64 bullous pemphigoid- proven patients (specifically IgG1, IgG3, IgG4), which were false-negative by classical indirect immunofourescence. We assessed the operating characteristics of an IgG subclass, which have not yet been determined.

We found out, that complement fixation test is suitable for the diagnosis of bullous pemphigoid and can help serologically challenging cases. Its´ sensitivity is 71,7 %, its´ specificity is 100 %. Assessment of Immunoglobulin G subclasses, especially IgG1 and IgG4 antibodies by indirect imunofluorescence on monkey esophagus can significantly improve diagnostic performance of indirect immunofluorescence in the bullous pemphigoid patients, the sensitivity of earlier false negative patients increased up to 48, 8 % with 97% specificity. In conclusion, our hypothesis is beeing confirmed.