Summary

Background: Heavy proteinuria may be caused by either increased glomerulal basement membrane permeability or membrane or podocyte structural damage, and also by impairment of secretion-reabsorption tubular processes. In this study, 60 patients with nephrotic proteinuria and other diagnoses (lupus nephritis, membranous nephropathy, IgA nephropathy, Wegener´s granulomatosis) and 20 patients with Anderson-Fabry disease (AFD),which is an X-linked genetic disorder with deficient *a*-galactosidase A activity, were analysed by the 2D electrophoresis method. The main aim of this work was to investigate possible differences in urine proteins in nephropaties, between healthy controls and AFD patients and to identify abnormal proteins as potential biomarkers of disease.

Methods: The urine proteins were devided by isoelectric focusing method using polyacrylamide strips (pH 3-10 linear). The second dimensional SDS electrophoresis was performed in 12 % polyacrylamide gel. The proteins were visualized by silver method and selected proteins were identified by MALDI-TOF MS. The gels were evaluated by Phoretix 2D expression software 2005.

Results: We found out that without adding protease inhibitors we can detect proteolysis, with increased quantity of proteins manifested in the area about 10 kDa and decreased quantity of proteins detectable in the area with molecular weights about 50 kDa. The separation of albumin caused higher lucidity of the urinary proteomes. The urinary maps comparison brought out that there are significant proteins´ changes, which are typical for Anderson-Fabry´s disease and other nephropathies and possible glycosylation at Asn51 and Asn78 sites of the prostaglandin H2 D-isomerase was detected in AFD patients. Also albumin, transferrin, alpha-1 antitrypsin and transthyretin precursor were identified by MALDI-TOF MS.

Conclusion: Changes of urinary proteins should be important for renal diagnosis and progression. AFD urinary proteomics revealed increased secretion of several proteins. We postulate that the observed difference in the amount of prostaglandin H2 D-isomerase and its position on two dimensional gels might be related to different glycosylation in AFD subjects.

Keywords: Anderson-Fabry disease/ MALDI-TOF MS/ two-dimensional electrophoresis/ urinary proteomes / nephrotic syndrome