Summary

Introduction: Idiopathic inflammatory myopathies (myositis, IIM) are heterogeneous group of rare autoimmune systemic diseases, characterized particularly by proximal skeletal muscle weakness. Heretogeneity of myositis is based on different pathogenetic mechanisms which may be reflected by variable imunophenotypic response in individual subtypes.

Objectives: The aim of this work was to explore the associations and influence of soluble factors of immune system in patient's sera on phenotypic characteristics and subtypes of IIM, to describe their expression in inflammed muscle tissue and study their eventual role in pathogenesis by analysis of effect on immune and muscle cells *in vitro*.

Results: We have described prevalence and characteristics of joint involvement in myositis patients and its significant association with anti-Jo-1 autoantibody. Further we confirmed the relation of anti-HMGCR antibody to immune mediated necrotizing myopathy, its tight relation to statins and recent increase in incidence. We showed inverse association of IFNα serum levels with muscle activity detected on MRI. Clinical activity positively correlated with IFN type-I pathway activation in patients with dermatomyositis. We also show positive correlation of resistin levels and clinical activity and correlation of activity with visfatin serum levels in anti-Jo-1 positive patients. Both resistin and visfatin are up-regulated in muscle biopsies. Moreover, we showed differentially expressed characteristic miRNA in sera of patients with PM and DM. Sera of patients with IIM are capable to activate IFN-type I pathway *in vitro* and IFNα seems to be responsible for that. We also demonstrate the ability of resistin to induce expression of pro-inflammatory cytokines (IL-1β, IL-l, MCP-1) in mononuclear cells.

Conclusions: Our results show the relation of particular molecules of immune system to individual subtypes of IIM and their phenotypic manifestations and suggest the role of soluble mediators in pathogenesis of idiopathic inflammatory myopathies.

Key words: idiopathic inflammatory myopathies, autoantibodies, cytokines