

## **Abstract**

This thesis includes set of published experimental results, which were obtained at the Department of Immunology, Second Faculty of Medicine Charles University within the project focused on the basal and applied research of the Primary immunodeficiencies (PID), particularly Common variable immunodeficiency (CVID).

The first, theoretical part, is divided into two sections. The first section is dedicated to the general aspects of Primary antibody deficiencies (PAD). The second section is focused on epidemiology, etiopathogenesis, classification, clinical and therapeutical aspects of CVID. The main consideration is devoted particularly to non-infectious complications, which significantly contribute to morbidity and mortality of CVID patients.

The second part consists of the set of publications describing specific mechanisms of immune system dysregulation and their clinical manifestation, which are briefly commented. The spectrum of issues resolved within the project covers following basic aspects: 1) characteristics of lung complications in CVID from the point of view of bronchial asthma, 2) characteristics of associated malignancies, 3) significance of genetic background for the specific treatment, 4) therapy of CVID focused on aspects of immunoglobulin substitution.

The results of the other experimental projects, which are not consistent with this thesis, and other publications including review articles or articles published in the Czech peer reviewed journals (without impact factor) are mentioned in the end in the chapter 12 „The list of publications“)

**Key words:** *Common variable immunodeficiency , bronchial asthma, genetics, malignancies, immunoglobulin substitution therapy*