

Investigators:

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Familial hemophagocytic lymphohistiocytosis (FHL) is a life-threatening primary immunodeficiency which is fatal without treatment. The disease is characterised by a severe disturbance of the immune homeostasis. Most cases of FHL are due to genetic defects in Perforin or in other genes involved in cellular cytotoxicity by NK cells or cytotoxic T cells. The hemophagocytic syndrome (HLH) frequently develops in the context of infections. The pathogenesis of this disease remains poorly understood. However, this is an important prerequisite for development of targeted immunological therapies. In this interdisciplinary project (collaboration with Prof. Stephan Ehl and Prof. Andreas Diefenbach) we want to further characterize the pathogenesis of FHL in animal models. The experiments address the following questions: 1. Which initial triggers are required for induction of HLH in FHL mice? 2. How does the nature of the genetic HLH defect influence the threshold for the induction of HLH?