Essential fatty acid (EFA) deficiency is a common condition in children with severe liver disease, especially in children with Cholestasis induced failure to thrive (CIFTT). The nutritional status of these children deteriorates due to the EFA deficiency. In order to improve the nutritional status of children with CIFTT, scientific research concerning the small intestinal function during EFA deficiency is required.

In our laboratory, a mouse model for EFA deficiency has been developed. Stable isotope dilution studies in this mouse model clearly show that EFA deficiency leads to a variety of functional changes in the small intestine. More specifically, lipid malabsorption and disaccharide digestion are impaired during EFA deficiency in mice. Increased intestinal reabsorption of bile salts is insufficient to normalize the decreased lipid absorption, underscoring previous implications that intracellular rather than intraluminal steps of fat absorption are impaired during EFA deficiency in mice. Short term supplementation of linoleic acid in vitro does not seem to reverse the effects of EFA deficiency on the small intestinal enterocytes.

Overall, the experiments described in this dissertation contribute to the understanding of the negative effects of EFA deficiency on the small intestinal function. Future patients studies using stable isotope-labeled macronutrients, i.e. lipids, carbohydrates and proteins, will further assess nutritional status of children with CIFTT. We expect that studies describe in this dissertation, along with nutritional studies in CIFTT patients, will help to design optimized nutritional formulas for children with CIFTT and will thereby improve the quality of life of these patients.