Huntington’s disease (HD) is an inherited autosomal dominant neurodegenerative disorder caused by an unstable expansion of the CAG repeat sequence within the huntingtin gene. Huntingtin associates with ubiquitin-proteasome system that ensures degradation of particular proteins including β-catenin which is an important molecule whose equilibrated degradation is necessary for the proper functioning of the Wnt signaling pathway. The binding of β-catenin to the destruction complex is altered in HD, leading to the toxic stabilization of β-catenin. The main goal of my thesis was to determine whether the accumulation of β-catenin due to the presence of mutant huntingtin is also characteristic of Liběchov minipigs, a large animal model of Huntington´s disease stably expressing N-truncated human mutant huntingtin. Using immunoblot and specific antibodies, we have revealed age-dependent accumulation of mutant huntingtin in transgenic minipigs. Unlike endogenous huntingtin, no decrease of the level of mutant huntingtin was observed in the striatum of transgenic animals. Surprisingly, this was followed by a decrease of phosphorylated β-catenin. Nevertheless, our results demostrate the accumulation of β-catenin in mesenchymal stem cells isolated from the oldest boars during ontogenesis. Furthermore, we have revealed a significant decrease of unphosphorylated β-catenin in some transgenic minipigs. Based on our findings, we assume that the phosphorylated β-catenin occurs predominantly in cytoplasm whereas the level of unphosphorylated β-catenin appeared to be increased in the nucleus of transgenic cells where it may regulate transcription of certain genes. Using proteomic methods, we were able to identify cytoskeletal proteins with strongly amplified phosphorylation in transgenic cells with profilin and vimentin playing a key role as the phosphorylation of these proteins is neccesary for initiation of mutant huntingtin aggregation.