SUMMARY

Hereditary cerebellar ataxias represent a heterogeneous group of neurodegenerative disorders. Variability of human hereditary ataxias is also reflected in animal models of cerebellar disorders. The most frequently used animal models are *Lurcher* and *Purkinje cell degeneration* (*pcd*) mutant mice.

The main aim of this thesis was to analyze and compare the spatial and emotional behavior of these cerebellar mutants. Additional aims were to study the impact of abnormal behavior on breeding capacity in *Lurcher* mice and to assess the applicability of cerebellar mutants as models for experimental therapy of cerebellar degeneration.

We have confirmed several behavioral impairments in both *Lurcher* and *pcd* mutant mice. Nevertheless, we have found that the manifestation of spatial behavior deficit is different in these two cerebellar mutants. Based on our findings, we propose that the deficit of spatial performance in cerebellar mutants may potentially arise from a combination of 1) cognitive disturbances, 2) sensory deficits, 3) motor impairments, and finally, 4) affective disorder. Moreover, resulting spatial behavior could also be modified by the specific effect of mutation, genetic background, and sex. We have also shown that abnormal behavior, e.g. maternal infanticide leads to decreased breeding capability in *Lurcher* females. Although we have shown that embryonic cerebellar grafts survive well in both *Lurcher* and SCA2 mice, the morphology of the graft did not promise any strong specific behavioral effects.