

R6/2 Transgenic Mouse Model

R6/2 mice express a N-terminal fragment of the Huntington's Disease (HD) gene under control of human gene promoter elements with >120 CAG repeats that are sufficient to produce the phenotype of HD:

- HTT aggregates
- Motor deficits
- Learning deficits
- Mean survival of appr. 100 days

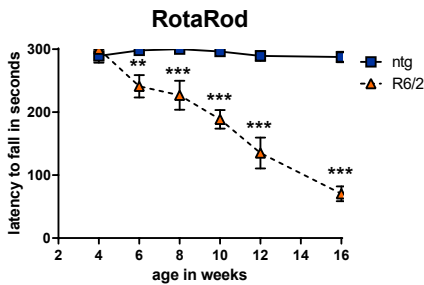


Figure 1: RotaRod test of R6/2 mice. Motor coordination expressed as time to fall off the rod in seconds of R6/2 and ntg mice. Mean $\pm$ SEM, Two-way ANOVA with Bonferroni's *post hoc* test; R6/2: n = 10, ntg: n = 19; **p<0.01; ***p<0.001.

Brain Atrophy

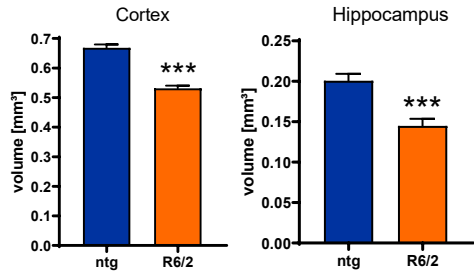


Figure 2: Brain atrophy in 4 month old R6/2 mice reflecting an approximately 20% grey matter loss in both investigated regions compared to age-matched ntg littermate. Mean $\pm$ SEM; unpaired t-test; n = 5; ***p<0.001.

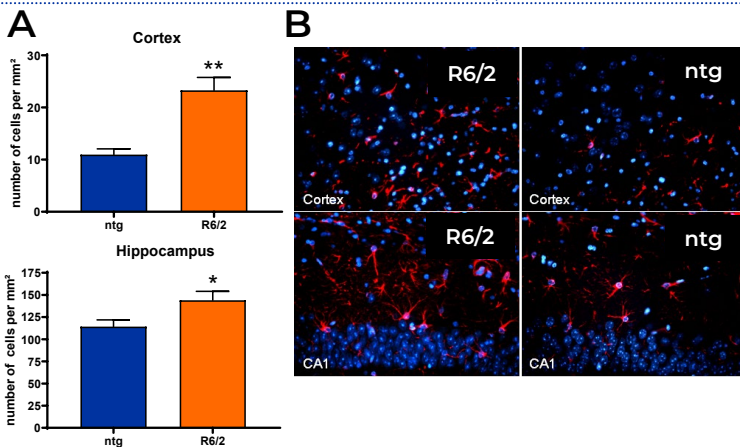


Figure 3. Astrogliosis in R6/2 mice. Quantitative and qualitative comparison of astrocytosis in R6/2 and non-transgenic (ntg) mice. A: Number of astrocytes per mm². Mean $\pm$ SEM; unpaired t-test; n = 5; *p<0.05; **p<0.01. B: Representative images of GFAP (red) in primary somatosens. cortex and hippocampal CA1 region. DAPI (blue).

Mangiarini L., Sathasivam K., Seller M., Cozens B., Harper A., Hetherington C., Lawton M., Trotter Y., Lehrsach H., Davies S.W., Bates G.P. Exon 1 of the HD Gene with an Expanded CAG Repeat Is Sufficient to Cause a Progressive Neurological Phenotype in Transgenic Mice. *Cell*, 1996, Vol. 87, 493–506.