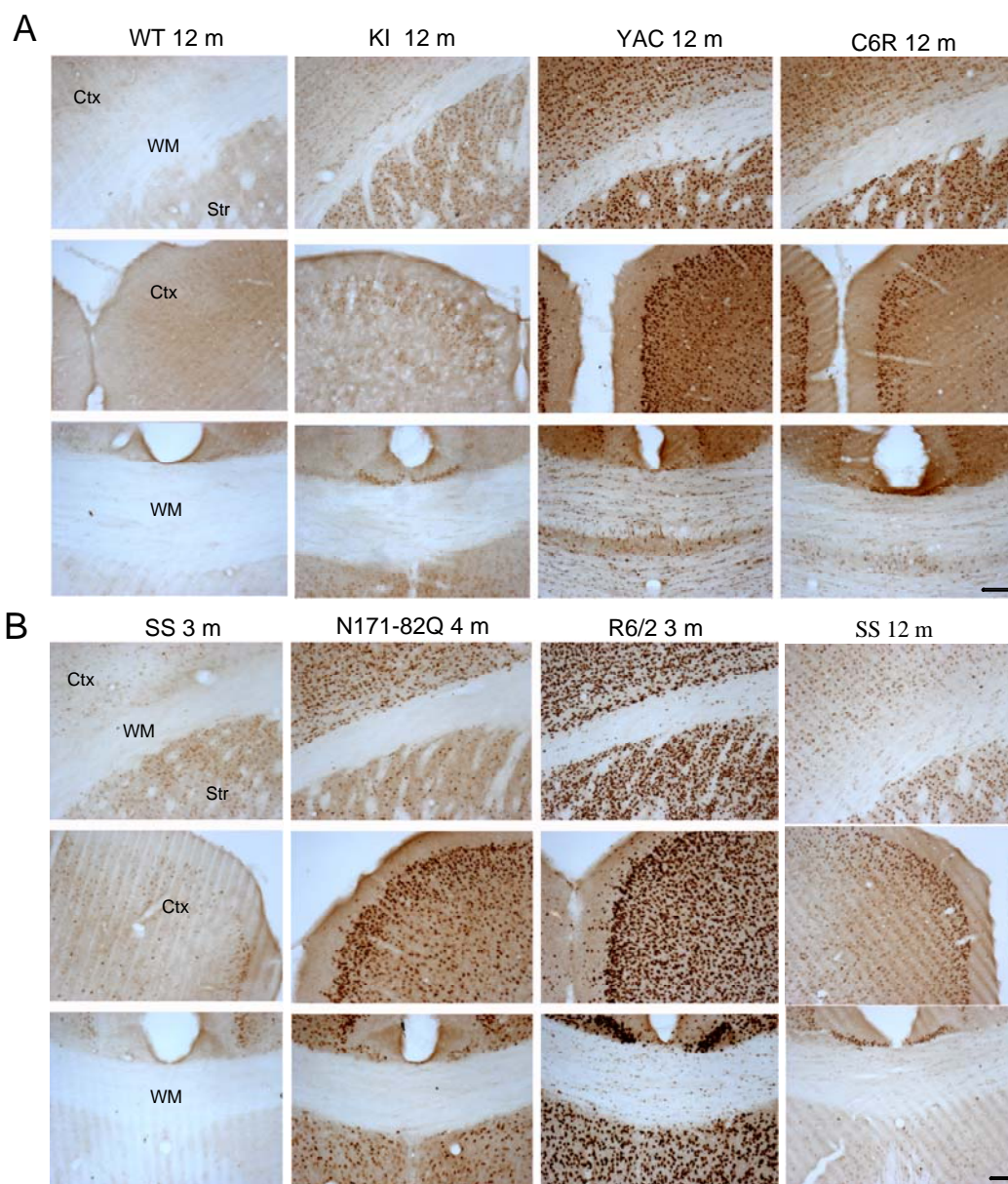


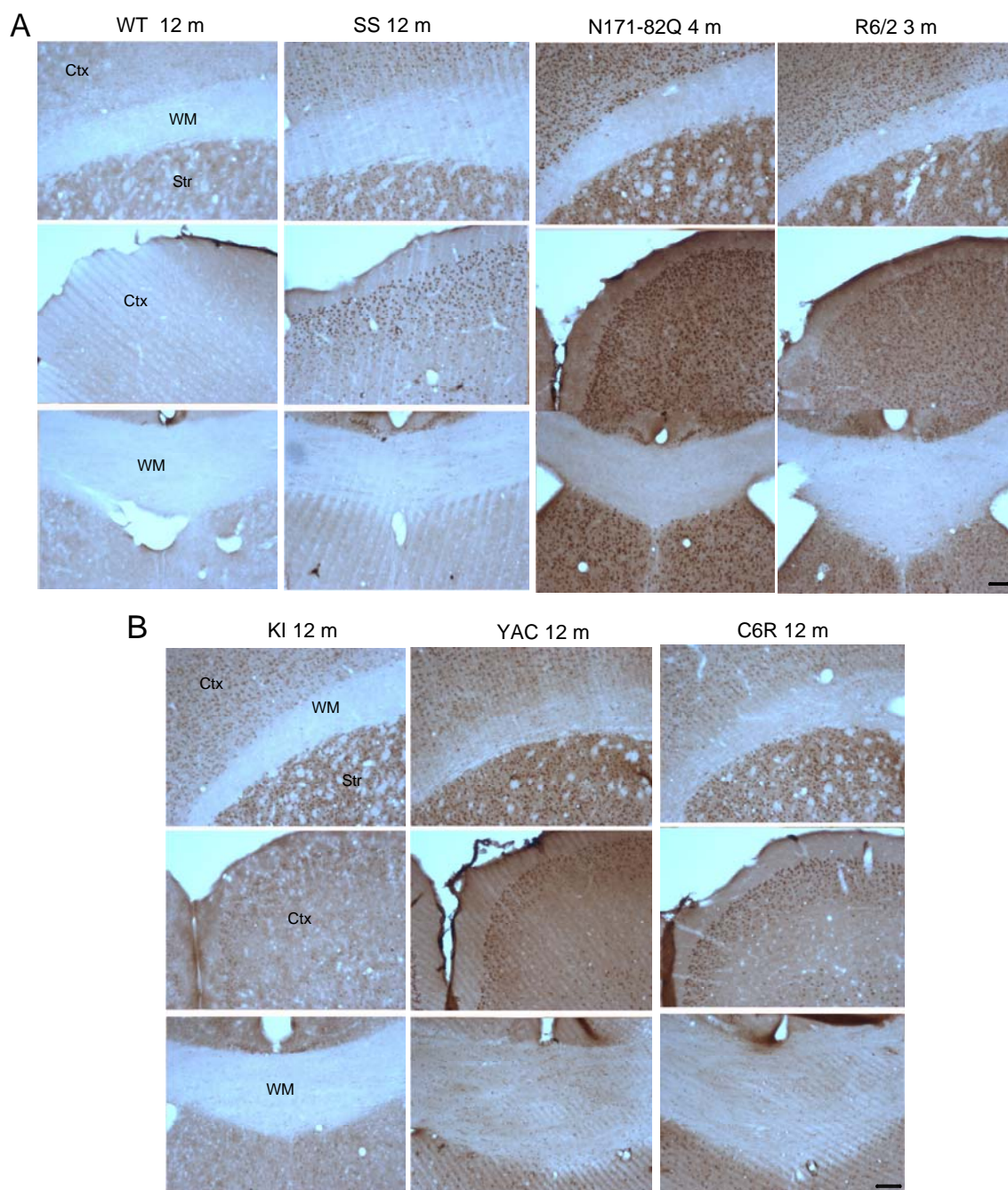
Supplemental figure 1. The sizes of the CAG repeat in various transgenic mouse models examined. (A) The sense primer 32S (5'-CTACGAGTCCCTCAAGTCCTTCCAGC-3') and the antisense primer 177A (5'-GACGCAGCAGCGGCTGTGCCTG-3') were used for PCR, which amplify the region encoding the CAG repeat. These primers are only able to amplify human htt sequences. (B) Agarose gel electrophoresis of PCR products from wild type (WT), YAC128, C6R, short stop (SS), N171-82Q, R6/2 mice. (C) The primers (OPD-8, 5'-CCCATTTCATTGCCTTGCTG-3' and HD 01-3, 5'-GCGGCTGAGGGGGTTGA-3'), which amplify mouse htt sequences, were used to detect the CAG repeat size in heterozygous (Het) and homozygous (Hom) HD150Q KI mice.

EM48 immunostaining



Supplemental figure 2. Low-magnification (10X objective lens) micrographs of EM48 immunohistochemistry. (A) EM48 staining of 12-month-old wild-type (WT) or HD mice expressing full-length mutant htt. KI: HD150Q knock-in; YAC: YAC128 transgenic mice; C6R: caspase-6-resistant YAC transgenic mice. (B) EM48 staining of transgenic mice expressing N-terminal mutant htt at the age of 3, 4, or 12 months as indicated. SS: shortstop transgenic mice; Ctx: cerebral cingulate cortex; Str: striatum; WM: white matter. The brain regions containing the striatum, white matter in the corpus callosum, cortex (upper panel), the 2nd motor cortex (middle panels), and white matter in the middle portion of the corpus callosum (bottom panels) are shown. Scale bars: 100 μ m.

1C2 immunostaining



Supplemental figure 3. Micrographs (10X objective lens) of 1C2

immunohistochemistry. (A) 1C2 staining of wild-type (WT) or transgenic mice expressing N-terminal mutant htt (B) at the age of 3, 4, or 12 months as indicated. (B) 1C2 staining of 12-month-old HD mice expressing full-length mutant htt. KI: HD150Q knock-in; YAC: YAC128 transgenic mice; C6R: caspase-6-resistant YAC transgenic mice. SS: shortstop transgenic mice; Ctx: cerebral cortex; Str: striatum; WM: white matter. The brain regions containing the striatum, white matter in the corpus callosum, cortex (upper panel), the 2nd motor cortex (middle panels), and white matter in the middle portion of the corpus callosum (bottom panels) are shown. Scale bars: 100 μ m.