



Supplementary Figure 4: Genomic organization of SCA8 region and *Klhl1* expression in BAC transgenic mice.

a) Comparison of the genomic organization of the overlapping *ATXN8* and *ATXN8OS* genes and 5'-region of the *KLHL1* gene. Exons are shown as boxes and various alternative splice forms are indicated by dashed lines. The murine *Klhl1as* transcript (a partial homolog of the much longer human *ATXN8OS* transcript) terminates within the *Klhl1* promoter region. The murine *Klhl1as* gene is much shorter than the human *ATXN8OS* transcript. Interestingly, the region of human genomic DNA containing *ATXN8OS* exons A, B, C1, C2, and C3 is not conserved in the downstream chromosomal region of the murine *Klhl1as* gene. **b)** Relative levels of murine *Klhl1* cerebellar RNA expression were assessed by Northern analysis using a 605 bp probe spanning exons2-5 of the murine *Klhl1* coding region. Relative expression levels in non-Tg (FVB), BAC-Exp1 and BAC-Ctl3 animals are comparable. **c)** Each bar represents an average of three animals with no significant differences between the BAC-Exp1 and FVB or BAC-Ctl3 animals.