

INVENTORY OF SUPPLEMENTAL INFORMATION:

Supplementary Figure legends

Figure S1. BAC fxAR121 mice express human AR protein in the CNS at levels comparable to endogenous mouse AR protein and human AR protein in the YAC AR100 mouse model. [related to Figure 1]

Figure S2. Male BAC fxAR121 mice exhibit muscle pathology and female BAC fxAR121 mice are normal. [related to Figure 2]

Figure S3. Cre-mediated reduction of hAR transgene expression in skeletal muscle does not alter endogenous mouse AR expression in BAC fxAR121 – HSA-Cre mice. [related to Figure 4]

Figure S4. Cre-mediated reduction of hAR transgene expression in skeletal muscle does not affect hAR expression in brain and does not prevent hAR protein aggregation in brain. [related to Figure 6]

Figure S5. Motor neuron axon numbers and calibers for control, BAC fxAR121, and BAC fxAR121 – HSA-Cre mice are comparable prior to onset of motor neuron degeneration. [related to Figure 7]