Mouse Models DRPLA Publication

Transgenic DRPLA 96Q Mice

IDENTIFICATION

Causal gene(s)	ATN1
Repeat size or mutation	96Q
Animal model	Mouse

MODEL DETAILS

Mouse strain / background	C57BL/6J
Type of model	Transgenic

TRANSGENIC CONSTRUCT

Transgenic construct: sequence type	Entire mutant DRPLA gene
Transgenic construct: details	Propagation of expansion through in vitro fertilization and transplantation creating a subline from the Q76 and Q129 mice
Promoter: gene	SV40

PHENOTYPE

Progression	Progressive
Hallmark features	Increased severity of motor deficits and memory disturbance with increasing CAG repeat length and age; reduced survival, reduced body weight, intranuclear inclusions, neuronal dysfunction

SOURCE & PUBLICATIONS

Originating lab / institution	The University of Tokyo
Links to publications or public resources	DRPLA transgenic mouse substrains carrying single copy of full-length mutant human DRPLA gene with variable sizes of expanded CAG repeats exhibit CAG repeat length- and age-dependent changes in behavioral abnormalities and gene expression profiles - PubMed https://pubmed.ncbi.nlm.nih.gov/22342974/