Mouse Models DRPLA Publication

Atro-118Q transgenic mice

IDENTIFICATION

Causal gene(s)	ATN1
Repeat size or mutation	118Q
Animal model	Mouse

MODEL DETAILS

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

TRANSGENIC CONSTRUCT

Transgenic construct: sequence type	Full length human cDNA
Transgenic construct: details	Full-length human atrophin-1 cDNA with 118Q was cloned into the NSE transgenic vector with a rat neuron-specific enolase promotor
Promoter: gene	NSE
Promoter: species	Rat

PHENOTYPE

Progression	Progressive
Hallmark features	Ataxia, tremors, motor defects, nuclear inclusions, somal atrophy, hypoacetylation of histone H3

SOURCE & PUBLICATIONS

Originating lab / institution	Morgan-Tan International Center for Life Sciences
Links to publications or public resources	Sodium butyrate ameliorates histone hypoacetylation and neurodegenerative phenotypes in a mouse model for DRPLA - PubMed https://pubmed.ncbi.nlm.nih.gov/16407196/