Mouse Models DRPLA Publication

AT-FL-65Q

IDENTIFICATION

Research Resource Identifier (RRID)	RRID: MMRRC_000398-MU
Causal gene(s)	ATN1
Repeat size or mutation	65Q
Animal model	Mouse

MODEL DETAILS

Mouse strain / background	C3H/B6
Type of model	Transgenic

TRANSGENIC CONSTRUCT

Transgenic construct: sequence type	Full-length cDNA
Transgenic construct: details	Full-length cDNA of atrophin-1 encoding 65Q constructs were ligated into the mouse prion protein promoter vector ensuring expression in neurons throughout the brain
Promoter: gene	PrP
Promoter: species	Mouse

PHENOTYPE

Progression	Progressive
Hallmark features	Involuntary tremors, ataxic gait, loss of coordination, hyperactivity, increased aggression, lower breeding efficiency, seizures, premature death, atrophin-1 nuclear inclusion bodies

SOURCE & PUBLICATIONS

Originating lab / institution	Johns Hopkins University School of Medicine
Links to publications or public resources	Nuclear accumulation of truncated atrophin-1 fragments in a transgenic mouse model of DRPLA - PubMed https://pubmed.ncbi.nlm.nih.gov/10677044/