Mouse Models FXTAS Publication

Gfa2-CGG99-eGFP mice

IDENTIFICATION

Causal gene(s)	FMR1
Repeat size or mutation	Human expanded CGG99 trinucleotide repeat segment
Animal model	Mouse

MODEL DETAILS

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

TRANSGENIC CONSTRUCT

Transgenic construct: sequence type	human expanded CGG99 trinucleotide repeat segment specifically in the astrocytes and Bergmann glia
Transgenic construct: details	Construct containing human expanded (CGG)99 and a GFP sequence under the Gfa2 promoter ensuring restricted expression in astrocytes and Bergmann glia
Promoter: gene	Gfa2
Promoter: species	Human

PHENOTYPE

Hallmark features	Impaired motor performance on ladder-rung test, improved motor performance on rotarod, intranuclear inclusions

SOURCE & PUBLICATIONS

Originating lab / institution	University of California
Links to publications or public resources	Astroglial-targeted expression of the fragile X CGG repeat premutation in mice yields RAN translation, motor deficits and possible evidence for cell-to-cell propagation of FXTAS pathology - PubMed https://pubmed.ncbi.nlm.nih.gov/30808398/