Mouse Models FXTAS Publication

TRE-90CGG-eGFP mice

IDENTIFICATION

Causal gene(s)	FMR1
Repeat size or mutation	Inducible transgenic mouse model that express 90 CGG
Animal model	Mouse

MODEL DETAILS

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

TRANSGENIC CONSTRUCT

Transgenic construct: details	Plasmid with eGFP with FMR1 5'UTR, 90CGGs under control of TRE (TRE-90CGG-eGFP) constructs were injected in mice oocytes. TRE-90CGG-eGFP mice were then crossed with the hnRNP-rtTA driver line
Promoter: gene	TRE and CMV

PHENOTYPE

Hallmark features	Weight loss, premature death, mitochondrial dysfunction, steatosis and apoptosis in the liver

SOURCE & PUBLICATIONS

Originating lab / institution	Erasmus Medical Center
Links to publications or public resources	Induced expression of expanded CGG RNA causes mitochondrial dysfunction in vivo - PubMed https://pubmed.ncbi.nlm.nih.gov/25486200/