B6J.129S1-*Htt* ^{tm1Mfc}/190ChdiJ

Stock No: **027410** | B6J.zQ175 KI ; Q175 Knock-In (wt x het) ; CHDI-81003003

Congenic, Targeted Mutation

Live mice available in varying quantities. Ask Customer Service for details.

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CAG repeat tract (see note below). These B6J.zQ175 KI mice (Stock No. 027410) are useful for studying Huntington's disease pathogenesis and for the assessment of potential therapeutic interventions.

This zQ175 KI allele is the same design as the zQ175 neo-deleted knock-in allele (zQ175DN KI; Stock No. 029928), with the exception that this zQ175 KI retains the floxed neo cassette. It is not specifically characterized if deletion of the neo cassette alters the zQ175DN KI phenotype compared to the zQ175 KI phenotype.

As of November 2017, the Stock No. 027410 live colony has 180-220 CAG repeats.

Donating Investigator

Marie-Francoise Chesselet, University of California, Los Angeles

Dr. David Howland, CHDI Foundation

Daniela Brunner, PsychoGenics Inc.

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GENETIC OVERVIEW

Genetic Background

Generation

?+N15
(2021-03-10 00:00:00)

Htt^{tm1Mfc}

Allele Type	Gene Symbol	Gene Name
Targeted (Humanized sequence)	*Htt*	huntingtin

VIEW GENETICS

RESEARCH APPLICATIONS

Neurobiology Research
Research Tools

VIEW ALL RESEARCH APPLICATION

BASE PRICE

Starting at:

$255.00 Domestic price for female 4-week

VIEW PRICE LIST

Details

Detailed Description

Stock No. 027410 was formerly associated with CHDI Foundation colony Stock No. 370476 [CHDI-81003003].

The phenotype description below is for zQ175 KI mice with ~190 CAG repeat length. Importantly, as of November 2017, the Stock No. 027410 live colony has 180-220 CAG repeats.
Huntington's disease (HD) is an autosomally dominant, fatal neurodegenerative disorder characterized by uncontrolled movements, psychiatric disturbances and cognitive impairment. HD is caused by an unstable trinucleotide (CAG) repeat expansion in the huntingtin gene (*HTT*; HD or Hdh).
The zQ175 knock-in (zQ175 KI) allele replaces mouse *Htt* exon 1 with the human *HTT* exon 1 sequence with ~190 repeats of a pure CAG tract [(CAG)nCAACAG, encoding polyglutamine]. The CAG repeat number is subject to germline and somatic instability, and may expand or contract. zQ175 mice have mutant mouse/human chimeric protein expression in brain at similar levels of normal endogenous huntingtin protein. Homozygous and heterozygous mice are viable and fertile with some characteristics that phenocopy Huntington's disease. Homozygous mice exhibit weakened grip strength (~4 weeks of age), motor deficit (~8 weeks of age), impaired rotarod and climbing activity (~30 weeks of age), circardian rhythm disruption (~9 months of age), cognitive deficits (~1 year of age), operant learning deficits (~1 year of age) and significantly reduced survival (median ~90 weeks of age). In addition, homozygous mice have huntingtin inclusions/aggregates (~2-4 months of age), early and significant decreased striatal gene markers (from ~12 weeks of age) and decreased neuronal cell counts. Heterozygous mice show behavioral deficits from around 4.5 months of age, especially in the dark phase of the diurnal cycle. Heterozygous mice also show motivational deficits (~30 weeks of age) and operant learning deficits (~1 year of age). Decreased expression of striatal gene markers are detected in heterozygous mice from ~18 weeks of age. Both heterozygotes and homozygotes have decreased body weight (from ~8 weeks of age).

This zQ175 KI allele (Stock No. 027410) is the same design as the zQ175 neo-deleted knock-in allele (zQ175DN KI; Stock No. 029928), with the exception that this zQ175 KI retains the floxed neo cassette. It is not specifically characterized if deletion of the neo cassette alters the zQ175DN KI phenotype compared to the zQ175 KI phenotype.

This Huntington's disease mouse model is available by way of a collaborative effort between CHDI Foundation, Dr. Scott Zeiltin (University of Virginia), Dr. Marie-Francoise Chesselet (University of California, Los Angeles), PsychoGenics, Inc. and The Jackson Laboratory.

In these mutant mice, the CAG repeat number is subject to germline and somatic instability, and may expand or contract. When using lines with unstable CAG repeat length, it is strongly recommended the CAG repeat number be quantified in all the experimental animals; all animals in all experimental groups should carry comparable CAG repeat sizes. CAG repeat sizing of HD mice should be done using high-resolution methods; as assays based on agarose gel

electrophoresis typically do not provide sufficient resolution to accurately measure CAG repeat numbers. If labs do not have access to the appropriate equipment for determining CAG repeat length, CAG repeats can be evaluated on a fee-for-service basis by Laragen, Inc.

- Development
- Control Suggestions
- Selected References

Genetics

- *Htt^{tm1Mfc}*

Disease/Phenotype

- Disease Terms
- Research Areas By Phenotype
- Mammalian Phenotype Terms by Genotype
- References

Technical Support

CONTACT TECHNICAL SUPPORT

Genotyping Protocols

Standard PCR:Laragen
Standard PCR:Laragen
QPCR:Generic Neo Quantitative PCR-QPCR- 1.2
Standard PCR:Generic Neo
Probe:Generic Neo
Genotyping resources and troubleshooting

Breeding Considerations

When maintaining our live colony, heterozygous mice are bred to C57BL/6J inbred mice (Stock No. 000664).

Additional Breeding and Husbandry Support

Mating System

C57BL/6J (000664) x Heterozygote
Heterozygote x C57BL/6J (000664)

Citation

When using the B6J.zQ175 KI ; Q175 Knock-In (wt x het) ; CHDI-81003003 mouse strain in a publication, please cite the originating article(s) and include JAX stock #027410 in your Materials and Methods section.

Animal Health Reports

Facility Barrier Level Descriptions

AX18 (Maximum)

Pricing & Availability

Available

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Domestic **International**

Pricing effective for USA, Canada and Mexico shipping destinations

LIVE MOUSE

AGE	SEX	GENOTYPE	PRICE
4 weeks	Female	Heterozygous for Htt^{tm1Mfc}	$255.00
	Male	Heterozygous for Htt^{tm1Mfc}	$255.00
4 weeks	Female	Wild-type for Htt^{tm1Mfc}	$78.51
	Male	Wild-type for Htt^{tm1Mfc}	$78.51
5 weeks	Female	Heterozygous for Htt^{tm1Mfc}	$255.00
	Male	Heterozygous for Htt^{tm1Mfc}	$255.00
5 weeks	Female	Wild-type for Htt^{tm1Mfc}	$78.51
	Male	Wild-type for Htt^{tm1Mfc}	$78.51
6 weeks	Female	Heterozygous for Htt^{tm1Mfc}	$255.00
	Male	Heterozygous for Htt^{tm1Mfc}	$255.00
6 weeks	Female	Wild-type for Htt^{tm1Mfc}	$78.51
	Male	Wild-type for Htt^{tm1Mfc}	$78.51
7 weeks	Female	Heterozygous for Htt^{tm1Mfc}	$255.00
	Male	Heterozygous for Htt^{tm1Mfc}	$255.00

	SEX		
7 weeks	Female	Wild-type for Htt^{tm1Mfc}	$78.51
	Male	Wild-type for Htt^{tm1Mfc}	$78.51
8 weeks	Female	Heterozygous for Htt^{tm1Mfc}	$255.00
	Male	Heterozygous for Htt^{tm1Mfc}	$255.00
8 weeks	Female	Wild-type for Htt^{tm1Mfc}	$78.51
	Male	Wild-type for Htt^{tm1Mfc}	$78.51
9 weeks	Female	Heterozygous for Htt^{tm1Mfc}	$255.00
	Male	Heterozygous for Htt^{tm1Mfc}	$255.00
9 weeks	Female	Wild-type for Htt^{tm1Mfc}	$78.51
	Male	Wild-type for Htt^{tm1Mfc}	$78.51
10 weeks	Female	Heterozygous for Htt^{tm1Mfc}	$255.00
	Male	Heterozygous for Htt^{tm1Mfc}	$255.00
10 weeks	Female	Wild-type for Htt^{tm1Mfc}	$78.51
	Male	Wild-type for Htt^{tm1Mfc}	$78.51
11 weeks	Female	Heterozygous for Htt^{tm1Mfc}	$255.00
	Male	Heterozygous for Htt^{tm1Mfc}	$255.00
11 weeks	Female	Wild-type for Htt^{tm1Mfc}	$78.51
	Male	Wild-type for Htt^{tm1Mfc}	$78.51
12 weeks	Female	Heterozygous for Htt^{tm1Mfc}	$255.00
	Male	Heterozygous for Htt^{tm1Mfc}	$255.00
12 weeks	Female	Wild-type for Htt^{tm1Mfc}	$78.51
	Male	Wild-type for Htt^{tm1Mfc}	$78.51

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