B6J.129S1-Htt<sup>9mi1Mc<sub>i</sub>/190ChdiJ</sup>

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

![Congenic Targeted Mutation]

AVAILABLE

PLACE ORDER

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

---

**Overview**

**Also Known As:** B6J.zQ175 KI ; Q175 Knock-In (wt x het) ; CHDI-81003003

The zQ175 knock-in (zQ175 KI) allele has the mouse Htt exon 1 replaced by the human HTT exon 1 sequence with a ~190 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.

GENETIC OVERVIEW

<table>
<thead>
<tr>
<th>Genetic Background</th>
<th>Generation</th>
</tr>
</thead>
<tbody>
<tr>
<td>?+N9</td>
<td>(2018-12-03 00:00:00)</td>
</tr>
</tbody>
</table>

Htt\textsuperscript{m1Mfc}

<table>
<thead>
<tr>
<th>Allele Type</th>
<th>Gene Symbol</th>
<th>Gene Name</th>
</tr>
</thead>
<tbody>
<tr>
<td>Targeted (Humanized sequence)</td>
<td>Htt</td>
<td>huntingtin</td>
</tr>
</tbody>
</table>

RESEARCH APPLICATIONS
Neurobiology Research
Research Tools

BASE PRICE
Starting at:
$76.22 Domestic price for female

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), circadian 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 Chesneau (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.
Genotyping Protocols
MELT: 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 B6;129S1-C192ICR(129-C192ICR)3.Aggemm(C192ICR)IvZ mouse strain in a publication, please cite the originating articles and include JAX stock #027410 in your Materials and Methods section.

Animal Health Reports

Facility Barrier Level Descriptions
AX18 (Maximum)

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

Domestic
International
Pricing effective for USA, Canada and Mexico shipping destinations

<table>
<thead>
<tr>
<th>Live Mouse</th>
<th>AGE</th>
<th>SEX</th>
<th>GENOTYPE</th>
<th>PRICE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Approx 4-8 weeks</td>
<td>Female</td>
<td>Heterozygous for Htt&lt;sup&gt;tm1Mfc&lt;/sup&gt;</td>
<td>$255.00</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Male</td>
<td>Heterozygous for Htt&lt;sup&gt;tm1Mfc&lt;/sup&gt;</td>
<td>$255.00</td>
<td></td>
</tr>
<tr>
<td>Approx 4-8 weeks</td>
<td>Female</td>
<td>Wild-type for Htt&lt;sup&gt;tm1Mfc&lt;/sup&gt;</td>
<td>$76.22</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Male</td>
<td>Wild-type for Htt&lt;sup&gt;tm1Mfc&lt;/sup&gt;</td>
<td>$76.22</td>
<td></td>
</tr>
</tbody>
</table>

Payment Terms and Conditions
Terms are granted by individual review and stated on the customer invoice(s) and account statement. These transactions are payable in U.S. currency within the granted terms. Payment for services, products, shipping containers, and shipping costs that are rendered
are expected within the payment terms indicated on the invoice or stated by contract. Invoices and account balances in arrears of stated terms may result in The Jackson Laboratory pursuing collection activities including but not limited to outside agencies and court filings.

The Jackson Laboratory’s Genotype Promise

The Jackson Laboratory has rigorous genetic quality control and mutant gene genotyping programs to ensure the genetic background of JAX® Mice strains as well as the genotypes of strains with identified molecular mutations. JAX® Mice strains are only made available to researchers after meeting our standards. However, the phenotype of each strain may not be fully characterized and/or captured in the strain data sheets. Therefore, we cannot guarantee a strain's phenotype will meet all expectations. To ensure that JAX® Mice will meet the needs of individual research projects or when requesting a strain that is new to your research, we suggest ordering and performing tests on a small number of mice to determine suitability for your particular project.

Terms Of Use

Terms of Use
General Terms and Conditions

QUESTIONS ABOUT TERMS OF USE

Additional Use Restrictions Apply
Use of MICE requires an agreement from genOway prior to shipping.

Licensing Information
Phone: 207-288-6470
Email: TechTran@jax.org

JAX® Mice, Products & Services Conditions of Use
‘MICE’ means mouse strains, their progeny derived by inbreeding or crossbreeding, unmodified derivatives from mouse strains or their progeny supplied by The Jackson Laboratory ("JACKSON"). ‘PRODUCT(S)’ means biological materials supplied by JACKSON, and their derivatives. ‘SERVICES’ means projects conducted by JACKSON for other parties that may include but are not limited to the use of MICE or PRODUCTS. ‘RECIPIENT’ means each recipient of MICE, PRODUCTS, or SERVICES provided by JACKSON including each institution, its employees and other researchers under its control. MICE or PRODUCTS shall not be: (i) used for any purpose other than internal research, (ii) sold or otherwise provided to any third party for any use, or (iii) provided to any agent or other third party to provide breeding or other services. Acceptance of MICE, PRODUCTS or SERVICES from JACKSON shall be deemed as agreement by RECIPIENT to these conditions, and departure from these conditions requires JACKSON’s prior written authorization.

No Warranty
MICE, PRODUCTS AND SERVICES ARE PROVIDED ‘AS IS’. JACKSON EXTENDS NO WARRANTIES OF ANY KIND, EITHER EXPRESS, IMPLIED, OR STATUTORY, WITH RESPECT TO MICE, PRODUCTS OR SERVICES, INCLUDING ANY IMPLIED WARRANTY OF MERCHANTABILITY OR FITNESS FOR A PARTICULAR PURPOSE, OR ANY WARRANTY OF NON-INFRINGEMENT OF ANY PATENT, TRADEMARK, OR OTHER INTELLECTUAL PROPERTY RIGHTS.

Credit for PRODUCTS or SERVICES
In case of dissatisfaction for a valid reason and claimed in writing by a purchaser within ninety (90) days of receipt of, PRODUCTS or SERVICES, JACKSON will, at its option, provide credit or replacement for the PRODUCT received or the SERVICES provided; JACKSON makes no other representations and this shall be the exclusive remedy of the purchaser. Please note specific policy for live mice.

Animal Care and Use for SERVICES
Consistent with the requirement for a written understanding regarding animal care and use, the JACKSON Animal Care and Use Committee will review the animal care and use protocol(s) associated with any SERVICES to be performed at JACKSON, and JACKSON shall have ultimate responsibility and authority for the care of animals while on site or in JACKSON custody.

No Liability
In no event shall JACKSON, its trustees, directors, officers, employees, and affiliates be liable for any causes of action or damages, including any direct, indirect, special, or consequential damages, arising out of the provision of MICE, PRODUCTS, or SERVICES, including economic damage or injury to property and lost profits, and including any damage arising from acts or negligence on the
part of JACKSON, its agents or employees. Unless prohibited by law, in purchasing or receiving MICE, PRODUCTS, or SERVICES from JACKSON, purchaser or recipient, or any party claiming by or through them, expressly releases and discharges JACKSON from all such causes of action or damages, and further agrees to defend and indemnify JACKSON from any costs or damages arising out of any third party claims.

MICE, PRODUCTS or SERVICES are to be used in a safe manner and in accordance with all applicable governmental rules and regulations.

The foregoing represents the General Terms and Conditions applicable to JACKSON’s MICE, PRODUCTS or SERVICES. In addition, special terms and conditions of sale of certain MICE, PRODUCTS, or SERVICES may be set forth separately in JACKSON web pages, catalogs, price lists, contracts, and/or other documents, and these special terms and conditions shall also govern the sale of these MICE, PRODUCTS and SERVICES by JACKSON, and by its licensees and distributors.

Acceptance of delivery of MICE, PRODUCTS or SERVICES shall be deemed agreement to these terms and conditions. No purchase order or other document transmitted by purchaser or recipient that may modify the terms and conditions hereof, shall be in any way binding on JACKSON, and instead the terms and conditions set forth herein, including any special terms and conditions set forth separately, shall govern the sale of MICE, PRODUCTS or SERVICES by JACKSON.

Related Strains

- All
- By Allele
- By Gene
- By Collection

All Related Strains

---

DO YOU NEED BALB/c MICE?

Rely on JAX to provide the models you need, when you need them.

CONTACT
Leading the search for

TOMORROW'S CURES