STOCK Mecp2 tm1.1Jtc/SchvJ


Targeted Mutation

CRYO RECOVERY

PLACE ORDER

Typically mice are recovered in 10-14 weeks. Contact Customer Service to place an order or for more information.

Also Known As: B6J:129S6.MeCP2 R168X

Mecp2 R168X is one of the most common MeCP2 mutations associated with Rett syndrome (RTT). Mecp2 R168X mice express a truncated R168X protein with a partial NLS and lacking the transcriptional repression domain (TRD) for interaction with corepressors. This RTT model is useful for testing highly robust behavioral paradigms in preclinical drug trials.

Donating Investigator
Laura R Schaevitz, Tufts University

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Genetic Background

<table>
<thead>
<tr>
<th>Genetic Background</th>
<th>Generation</th>
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<tbody>
<tr>
<td>Mecp2 tm1.1Jtc</td>
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<table>
<thead>
<tr>
<th>Allele Type</th>
<th>Gene Symbol</th>
<th>Gene Name</th>
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<tr>
<td>Targeted (Humanized sequence)</td>
<td>Mecp2</td>
<td>methyl CpG binding protein 2</td>
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VIEW GENETICS

Neurobiology Research
Developmental Biology Research

VIEW ALL RESEARCH APPLICATIONS
The early truncating MeCP2\textsuperscript{R168X} nonsense point mutation in the methyl-CpG binding domain of MeCP2 is one of the most common MeCP2 mutations associated with Rett syndrome (RTT). The largely truncated R168X protein retains the capacity to bind methylated DNA and carries a partial nuclear localization signal, but lacks the transcriptional repression domain for interaction with corepressors. A second termination (BspH1 restriction site) downstream of the R168X mutation is reported not to affect the mutant product. MeCP2\textsuperscript{R168X} mice are a model of RTT, exhibiting respiratory, neuromuscular and behavioral abnormalities similar, but not identical, to that of Mecp2 null mice. The phenotype is described in detail below. These mice also underscore the importance of including Mecp2 mutant females in preclinical studies.

MeCP2\textsuperscript{R168X} mice on a mixed genetic background of -87-93% C57BL/6J and -13-6% 129S6/SvEvTac (B6.J:129S6.Mecp2\textsuperscript{R168X}) are Stock No. 024990. In an attempt to offer alleles on well-characterized or multiple genetic backgrounds, alleles are frequently moved to a genetic background different from that on which an allele was first characterized. It should be noted that the phenotype of these B6.J:129S6.Mecp2\textsuperscript{R168X} mice could vary from that originally described on other genetic backgrounds. We will modify the strain description if necessary as published results become available.

Lawson-Yeun et al. 2007 Brain Res 1180:1 reports the phenotype of Mecp2\textsuperscript{R168X} animals backcrossed at least 10 generation onto the 129S6/SvEvTac genetic background (129S6.Mecp2\textsuperscript{R168X}) as: hemizygous males (Mecp2\textsuperscript{R168X/Y}) have a shortened lifespan (average 86 days) with forelimb stereotypes, hindlimb atrophy, hypoactivity and breathing irregularities. By 7 weeks of age, significant hindlimb clamping is evident. Female heterozygotes (Mecp2\textsuperscript{R168X/+}) manifest significant defects by approximately 6 months (hindlimb clamping, breathing irregularities) and can survive past 1 year of age.

Schaevitz et al. 2013 Genes Brain Behav 12:732 reports the phenotype of 129S6.Mecp2\textsuperscript{R168X} animals backcrossed 1-2 generations onto C57BL/6J as: MeCP2\textsuperscript{R168X} mutants mirror many clinical features of human RTT. MeCP2\textsuperscript{R168X/Y} males exhibit growth, motor, respiratory and cognitive abnormalities, and reduced anxiety. The phenotype is less severe and with later onset in MeCP2\textsuperscript{R168X/+} females with the exception of seizures: ~4% of MeCP2\textsuperscript{R168X/+} females exhibit tonic-clonic seizures that typically result in death. The phenotype in MeCP2\textsuperscript{R168X/Y} males is similar to that reported for Mecp2 null mice (such as Mecp2\textsuperscript{tm11Bird}, Stock No. 003890). Compared to females heterozygous for the Mecp2 null mutation, MeCP2\textsuperscript{R168X/+} females exhibit delayed motor defect onset, normal anxiety-like behavior and increased seizure susceptibility.

Bissonnette et al. 2014 Neuroscience 267:166 reports the phenotype of Mecp2\textsuperscript{R168X} animals on a mixed C57BL/6J:129S6/SvEvTac genetic background as: Mecp2\textsuperscript{R168X/+} females display augmented hypoxic ventilatory responses and depressed hypercapnic responses; the incidence of apnea is much greater in Mecp2\textsuperscript{R168X/+} females (189 per hour) than Mecp2\textsuperscript{T158A/+} females (41 per hour).

For B6.J:129S6.Mecp2\textsuperscript{R168X} mice, the donating investigator reports that mating hemizygous males with wildtype females does not produce offspring; it is not known if the hemizygous males are sterile or if they simply do not breed as a result of their Rett syndrome-like phenotype. The donating investigator also reports that backcrossing MeCP2\textsuperscript{R168X} mice onto the C57BL/6 genetic background leads to increased phenotype severity.

The phenotype differences observed between male and female mice is because Mecp2 is located on the X chromosome. Due to X-chromosome inactivation, heterozygous females have mosaic expression of wildtype MeCP2.
Genotyping Protocols
Sanger sequencing: Mecp21tm1.Jlc
Genotyping resources and troubleshooting

Breeding Considerations
The MeCP21R168X mutant allele is located on the X chromosome. Hemizygous males exhibit features of Rett syndrome and have a shortened lifespan. Heterozygous females may live more than one year and have a less severe phenotype with later onset. When maintaining a live colony, heterozygous females may be bred with wildtype males from the colony. The donating investigator reports that mating hemizygous males with wildtype females does not produce offspring; it is not known if the hemizygous males are sterile or if they simply do not breed as a result of their Rett syndrome-like phenotype. The donating investigator also reports that backcrossing MeCP21R168X mice onto the C57BL/6 genetic background leads to increased phenotype severity. The expected coat colors are agouti and black.
Additional Breeding and Husbandry Support

Citation
When using the B6J-129S6-Mecp21R168X mouse strain in a publication, please cite the originating article(s) and include JAX stock #024990 in your Materials and Methods section.
Facility Barrier Level Descriptions
Production of mice from cryopreserved embryos or sperm occurs in a maximum barrier room, G200
Pricing & Availability

Cryo Recovery
Typically mice are recovered in 10-14 weeks. Contact Customer Service to place an order or for more information.

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

<table>
<thead>
<tr>
<th>Cryorecovery - Domestic Pricing</th>
<th>GENOTYPE</th>
<th>PRICE</th>
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<tr>
<td>Cryo Recovery</td>
<td>X linked = Females are heterozygous and males are wildtype for Mecp2&lt;tm1.1Jtc&gt; X linked</td>
<td>$2,595.00</td>
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We will fulfill your order by providing at least two carriers for each strain ordered. The total number, sex, and genotypes provided will vary, although typically 8 or more animals are provided. Please check genotypes which will be recovered. While the genotypes of all animals produced will be communicated to you prior to scheduling shipment, the genotypes of animals provided may not reflect the mating scheme and genotypes described in the strain description. Animals are typically ready to ship in 11-14 weeks. If a second recovery is required to produce the minimum number of animals, then delivery time would increase to approximately 25 weeks. If we fail to produce animals of the correct genotype, you will not be charged. We cannot guarantee the reproductive success of mice shipped to your facility. If the mice are lost after the first three days (post-arrival) or do not produce progeny at your facility, a new order and fee will be necessary.

Cryo recovery to establish a Dedicated Supply for greater quantities of mice. Mice recovered can be used to establish a dedicated colony to contractually supply you mice according to your requirements. Price by quotation.

Related Products and Services

| Frozen Mouse Embryo | $2,595.00 per straw or vial |

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