**B6SJL-Tg(Prnp-Immt/SOD1*G93A)7Gmnf/J**

**Stock No:** 025403 | mito-G93ASOD1

- Transgenic

[CRYO RECOVERY]

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

**Also Known As: mito-G93ASOD1**

mito-G93ASOD1 transgenic mice express mitochondrial inner membrane-tethered, human mutant SOD1<sup>G93A</sup> at high levels in brain, spinal cord, heart and skeletal muscle. Homozygous mito-G93ASOD1 mice exhibit progressive mitochondrial dysfunction and neurodegeneration, but no muscle denervation. These mice may be useful for studying the pathogenic role of SOD1 and mitochondria in familial amyotrophic lateral sclerosis (ALS or Lou Gehrig's disease).

**Donating Investigator**

Giovanni Manfredi, Weill Medical College of Cornell University

**GENETIC OVERVIEW**

<table>
<thead>
<tr>
<th>Genetic Background</th>
<th>Generation</th>
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<tr>
<td>Tg(Prnp-Immt/SOD1*G93A)7Gmnf</td>
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**Allele Type**

Transgenic (Inserted expressed sequence, Humanized sequence)

[VIEW GENETICS]

**RESEARCH APPLICATIONS**

- Research Tools
- Neurobiology Research
- Cell Biology Research

[VIEW ALL RESEARCH APPLICATIONS]
mito-G93ASOD1 transgenic mice have the mouse prion protein promoter directing expression of mutant human SOD1<sup>G93A</sup> with N-terminal fusion of the mouse mitofilin mitochondrial targeting signal, MMP cleavage site and transmembrane domain. This results in expression of SOD1<sup>G93A</sup> that is anchored to the outer side of the mitochondrial inner membrane (i.e., facing the intermembrane space) at high levels in brain, spinal cord, heart and skeletal muscle. High expression is observed in both neurons and astrocytes. Low to undetectable levels of protein are reported in kidney, lung, spleen and liver. The mito-G93ASOD1 protein in high expressing tissues oligomerizes and acquires enzymatic activity. Homozygous mito-G93ASOD1 mice are viable and fertile with central nervous system mitochondria defects but no overt neuromuscular abnormalities. Homozygous mito-G93ASOD1 mice exhibit mitochondrial dysfunction and neurodegeneration that results in morbidity/death by one year. Specifically, homozygotes develop a progressive disease characterized by body weight loss (by 8 months in females), muscle weakness (by 8 months in both sexes), brain atrophy (by 8 months in males), and motor impairment (by 3 months in females; by 6 months in males). The phenotype is more severe in females. These symptoms are associated with reduced spinal motor neuron counts and impaired mitochondrial bioenergetics, characterized by decreased cytochrome oxidase activity and defective calcium handling (by 12 months of age). Importantly, homozygous mito-G93ASOD1 mice show no evidence of muscle denervation (a major pathological feature of ALS).

Of note, homozygous mito-G93ASOD1 accumulate mitochondrial SOD1<sup>G93A</sup> at levels comparable to those found in the high-expressor hemizygous B6SJL-Tg-(SOD1<sup>G93A</sup>)1Gur/J mice (Stock No. 002726).

The mito-WTSOD1 transgenic mice (Stock No. 024502) are a control strain for mito-G93ASOD1 mice.
Mammalian Phenotype Terms by Genotype

References

Technical Support

Genotyping Protocols
High Resolution Melting: Tg(Prnp-Immt/SOD1)7Gmnf
Genotyping resources and troubleshooting

Breeding Considerations
When maintaining a live colony, hemizygous mice are bred to B6SJLF1/J mice (Stock No. 100012) every generation.
Additional Breeding and Husbandry Support

Mating System
Hemizygote x B6SJLF1/J (100012) and reci

Citation
When using the mito-G93ASOD1 mouse strain in a publication, please cite the originating article(s) and include JAX stock #025403 in your Materials and Methods section.

Animal Health Reports
Facility Barrier Level Descriptions
Production of mice from cryopreserved embryos or sperm occurs in a maximum barrier room, G200

Pricing & Availability

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

Cryorecovery - Domestic Not-For-Profit & Academic Pricing

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<tr>
<th>SERVICE</th>
<th>GENOTYPE</th>
<th>PRICE</th>
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<tbody>
<tr>
<td>Cryo Recovery</td>
<td>Hemizygous or Non carrier for Tg(Prnp-Immt/SOD1*G93A)7Gmnf</td>
<td>$2,595.00</td>
</tr>
</tbody>
</table>

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.

Cryorecovery 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.

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<tr>
<th>Related Products and Services</th>
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<tbody>
<tr>
<td>Frozen Mouse Embryo</td>
<td>$2,595.00 per straw or vial</td>
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