The Jackson Laboratory

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

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

Transgenic

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

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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

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GENETIC OVERVIEW

Genetic Background **Generation**

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

Allele Type

Transgenic (Inserted expressed sequence, Humanized sequence)

VIEW GENETICS

RESEARCH APPLICATIONS

Research Tools
Neurobiology Research
Cell Biology Research

VIEW ALL RESEARCH APPLICATIO

BASE PRICE

Starting at:

$2,854.50 Domestic price Cryo Recovery

VIEW PRICE LIST

Details

Detailed Description

mito-G93ASOD1 transgenic mice have the mouse prion protein promoter directing expression of mutant human *SOD1*^{G93A} with N-terminal fusion of the mouse mitofilin mitochondrial targeting signal, MMP cleavage site and transmembrane domain. This results in expression of SOD1^{G93A} 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. Hemizygous 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^{G93A} at levels comparable to those found in the high-expressor hemizygous B6SJL-Tg-(SOD1*G93A)1Gur/J mice (Stock No. 002726).

The mito-WTSOD1 transgenic mice (Stock No. 024502) are a control strain for mito-G93ASOD1 mice.

Development

Expression Data

Control Suggestions

Selected References

Genetics

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

Disease/Phenotype

Disease Terms

Research Areas By Phenotype

Mammalian Phenotype Terms by Genotype

References

Technical Support

CONTACT TECHNICAL SUPPORT

Genotyping Protocols

Standard PCR:Tg(Prnp-Immt/SOD1)1Gmnf
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

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

CRYORECOVERY - DOMESTIC NOT-FOR-PROFIT & ACADEMIC PRICING		
SERVICE/PRODUCT	DESCRIPTION	PRICE
Cryo Recovery	Hemiozygous or Non carrier for Tg(Prnp-Immt/SOD1*G93A)7Gmnf	$2,854.50

RELATED PRODUCTS AND SERVICES		
Frozen Mouse Embryo	B6SJL-Tg(Prnp-Immt/SOD1*G93A)7Gmnf/J Frozen Embryo	$2595.00

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. We do not guarantee breeding performance and therefore suggest that investigators order more than one breeding pair to avoid delays in their research.

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Email: TechTran@jax.org

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By Gene

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