MOUSE STRAIN DATASHEET - 005024

FVB.Cg-Grm7 Tg(SMN2)89Ahmb Smn1 tm1Msd J

Stock No: 005024 | FVB.SMN2;Smn- , Severe Type I SMA , Burghes' Severe Mode...

Congenic, Targeted Mutation, Transgenic

AVAILABLE

PLACE ORDER

Live mice available in varying quantities. Ask Customer Service for details.
Also Known As: FVB.SM2;Smn-, Severe Type I SMA, Burghes' Severe Model incipient congenic

Mice that are homozygous for the targeted mutant Smn1 allele and carry the SMN2 transgene exhibit symptoms and neuropathology similar to patients afflicted with type I proximal spinal muscular atrophy (SMA). Mice that survive for several days exhibit decreased suckling and movement, labored breathing and trembling limbs after 48 hours. Stillborn mice and mice that succumb later are noticeably smaller than normal littermates. Muscle fibers (quadriceps and gastrocnemius) in affected mice display atrophy.

Donating Investigator

Arthur H.M. Burghes, The Ohio State University
Mice that are homozygous for the targeted mutant Smn1 allele and carry the SMN2 transgene exhibit symptoms and neuropathology similar to patients afflicted with type I proximal spinal muscular atrophy (SMA). In the initial characterization by the donating investigator, mice were either stillborn or survived 4-6 days. Mice that died at or shortly after birth were slightly smaller (1.33 g. vs. 1.51 g.) than normal littermates. Mice that survive for several days are indistinguishable from normal littermates in the first 48 hours, after which they exhibit decreased suckling and movement, labored breathing and tremoring limbs. Mice succumbing at this later time point are noticeably smaller than normal littermates (1.47 g vs. 4.59 g). A bell-shaped trunk is also noticeable in affected mice, presumably from intercostal muscle weakness, a characteristic of type I SMA. Histological analysis indicates that affected mice that survive to day 5 exhibit a loss of motor neurons from spinal cord (35%) and facial nucleus (40%). A large number of cells with pyknotic nuclei are observed in these tissues. Immunohistochemical analysis indicates low-level expression of the SMN2 protein in the tissues examined (brain, liver, spinal cord) and an absence or near absence of intranuclear aggregates of the SMN protein ('gems'). The donating investigator reports that muscle fibers (quadriceps and gastrocnemius assayed) are atrophied, a characteristic observed in SMA patients. Homozygous mice bearing the Smn1 targeted mutation without a copy of the SMN2 transgene display an embryonic lethal phenotype with developmental arrest occurring prior to implantation.

Note: In contrast to the original publication, and possibly due to inbreeding, it is the experience at The Jackson Laboratory Repository that mice that are hemizygous for the Tg(SMN2)89 transgene and homozygous for the Smn1 null allele do not survive.

Importation of this model was supported by the Spinal Muscular Atrophy Foundation. Creation and development was supported by the National Institutes of Health, the Deutsche Forschungsgemeinschaft to M.S., Families of SMA, the Preston fund, the Madison fund, the Mathew fund and the Muscular Dystrophy Association of America.
Genotyping Protocols
QPCR: Grm7<sup>Tg(SMN2)89Ahmb<sup> tm1Msd</sup>
Standard PCR: Smn1<sup>tm1Msd</sup><br>Standard PCR: Grm7<sup>Tg(SMN2)89Ahmb</sup><br>Standard PCR: Smn1<sup>tm1Msd</sup><br>Separated PCR: Smn1<sup>Tg(SMN2)89Ahmb</sup><br>Standard PCR: Grm7

Genotyping resources and troubleshooting

Dietary Information
LabDiet® 5K52 formulation (6% fat)

Breeding Considerations
The Tg(SMN2)89 transgene insertion into the glutamate receptor metabotropic 7 locus (Grm7<sup>Tg(SMN2)89Ahmb</sup>) on chromosome 6 and the SMN null allele (Smn1<sup>tm1Msd</sup>) on chromosome 13 are not linked and will segregate independently. Breeding pairs offered by The Jackson Laboratory are homozygous for the transgene and heterozygous for the targeted Smn1 allele. These breeding pairs are phenotypically normal and do not exhibit symptoms of neuropathology. Offspring resulting from the mating of breeder pairs can possess the following genotypes:

1. Homozygous for the transgene and homozygous for the targeted mutation (25%)
2. Homozygous for the transgene and heterozygous for the targeted mutation (50%)
3. Homozygous for the transgene and wildtype at the Smn1 locus (25%)

Mice that are homozygous for the transgene and homozygous for the targeted mutation will display the SMA-like phenotype. Mice homozygous for the transgene and heterozygous for the targeted mutation will not display the SMA-like phenotype but can be mated with each other to generate additional affected mice. Mice homozygous for the transgene and wildtype at the Smn1 locus will also not exhibit an SMA-like phenotype but can be employed as control mice depending on the nature of the experiment. The Jackson Laboratory also distributes mice that are homozygous for the transgene and wildtype at the Smn1 locus.

Note: In contrast to the original publication, and possibly due to inbreeding, it is the experience at The Jackson Laboratory Repository that mice that are hemizygous for the Tg(SMN2)89 transgene and homozygous for the Smn1 null allele do not survive.

Additional Breeding and Husbandry Support
Mating System
See "Breeding Considerations"

Citation
When using the FVB.SMN2;Smn<sup>-</sup>, Severe Type I SMA, Sevugh's Severe Model incipient congenic mouse strain in a publication, please cite the originating article(s) and include JAX stock #005024 in your Materials and Methods section.

Animal Health Reports
Facility Barrier Level Descriptions
Live mice available in varying quantities. Ask Customer Service for details.

### Domestic

Pricing effective for USA, Canada and Mexico shipping destinations

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### International

Pricing & Availability


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### Notes

- All prices are in USD.
- Shipping costs are not included in the listed prices.
- Please contact Customer Service for detailed instructions and availability.
### Breeder Pair

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- By Collection

All Related Strains