Mice that are homozygous for the \textit{Smn} targeted mutant \textit{lacZ} reporter allele and homozygous for the two transgenic alleles exhibit symptoms and neuropathology similar to patients afflicted with proximal spinal muscular atrophy (SMA). Triple homozygous mice on the FVB/N genetic background are noticeably smaller than normal littermates at birth and show progressive muscle weakness.

Our preclinical efficacy testing services offer scientific expertise and an array of target-based and phenotype-based outcome measures, both in vivo and at endpoint, for flexible study designs and assay development in mouse models of Spinal Muscular Atrophy. \textit{See our full service platform.}

Donating Investigator

Arthur H.M. Burghes, The Ohio State University

**GENETIC OVERVIEW**

<table>
<thead>
<tr>
<th>Genetic Background</th>
<th>Generation</th>
</tr>
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<tbody>
<tr>
<td>FVB.Cg-Grm7 \textit{Tg(SMN2)8Ahmb} Smn1 \textit{tm1Msd} Tg(SMN2*delta7)4299Ahmb/J</td>
<td>\textit{N6+F39} (2019-12-19 00:00:00)</td>
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</tbody>
</table>

\textbf{Tg(SMN2*delta7)4299Ahmb}

\textbf{Allele Type}

Transgenic (Inserted expressed sequence, Humanized sequence)

\textbf{Grm7}\textit{Tg(SMN2)89Ahmb}

\textbf{Allele Type}

Transgenic (Hypomorph, Inserted expressed sequence, Humanized sequence)

\textbf{Smn1}\textit{tm1Msd}

<table>
<thead>
<tr>
<th>Allele Type</th>
<th>Gene Symbol</th>
<th>Gene Name</th>
</tr>
</thead>
<tbody>
<tr>
<td>Targeted (Reporter, Null/Knockout)</td>
<td>\textit{Smn1}</td>
<td>survival motor neuron 1</td>
</tr>
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</table>
This triple mutant mouse harbors two transgenic alleles and a single targeted mutant. The Tg(SMN2*delta7)4299Ahmb allele consists of a SMA cDNA lacking exon 7 whereas the Tg(SMN2)89Ahmb allele consists of the entire human SMN2 gene. Mice that are homozygous for the targeted mutant Smn allele and homozygous for the two transgenic alleles exhibit symptoms and neuropathology similar to patients afflicted with proximal spinal muscular atrophy (SMA). At birth, triple mutants are noticeably smaller than normal littermates. By day 5, signs of muscle weakness are apparent and become progressively more pronounced over the following week as the mice display an abnormal gait, shakiness in the hind limbs and a tendency to fall over. In 2006, mean survival was originally reported to be ~13 days. Cohorts tested at The Jackson Laboratory in 2013 show death by ~15-22 days (mean survival of 17.7 days).

Immunocytochemical analysis indicates that dystrophin expression is normal, however fibers isolated from the gastrocnemius muscle of a 14 day old triple mutant clearly show evidence of atrophy.

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.
Genetics

- Tg(SMN2*delta7)4299Ahmb
- Grm7Tg(SMN2)89Ahmb
- Smn1tm1Msd

Disease/Phenotype

- Disease Terms
- Research Areas By Phenotype
- Mammalian Phenotype Terms by Genotype
- References

Technical Support
Genotyping Protocols
QPCR: Tg(SMN2)
Standard PCR: Smn1
Standard PCR: Grm7
Standard PCR: Tg(SMN2*)
Standard PCR: Smn1
QPCR: Tg(SMN2*)
Probe: Grm7-Probe

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 (Grm7Tg(SMN2)89Ahmb) on chromosome 6, the Smn1 null targeted mutation (Smn1tm7Msd) on chromosome 13 and the randomly inserted Tg(SMN2*delta7)4299Ahmb transgene are not linked and will segregate independently.

Breeding pairs offered by The Jackson Laboratory Repository are homozygous for both transgenes (Δ7SMN*;SMN2*) and either heterozygous for the null allele (Smn*) or wildtype at the Smn1 locus (Smn**). 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 both transgenes and heterozygous for the null allele (Δ7SMN*;SMN2*;Smn* - 50%)
2. Homozygous for both transgenes and wildtype at the Smn1 locus (Δ7SMN*;SMN2*;Smn** - 50%)

Mice that are homozygous for the Smn1 null allele, Tg(SMN2)89 and Tg(SMN2*delta7)4299Ahmb will display the SMA-like phenotype. Mice heterozygous for the Smn1 null allele and homozygous for Tg(SMN2)89 and Tg(SMN2*delta7)4299Ahmb will not display the SMA-like phenotype - but can be mated with each other to generate additional affected mice. Mice wildtype at the Smn1 locus and homozygous for Tg(SMN2)89 and Tg(SMN2*delta7)4299Ahmb will also not exhibit an SMA-like phenotype - but can be employed as control mice depending on the nature of the experiment.

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

Citation

When using the FVB.SMNΔ7;SMN2;Smn- , Moderate Type II SMA , Delta 7 mouse incipient congenic mouse strain in a publication, please cite the originating article(s) and include JAX stock #005025 in your Materials and Methods section.

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Facility Barrier Level Descriptions

- AX11 (Maximum)

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International

Pricing effective for USA, Canada and Mexico shipping destinations

LIVE MOUSE
<table>
<thead>
<tr>
<th>AGE</th>
<th>SEX</th>
<th>GENOTYPE</th>
<th>PRICE</th>
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<td>$270.00</td>
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<tr>
<td>4 weeks</td>
<td>Male</td>
<td>Homozygous for Grm7&lt;sub&gt;Tg(SMN2<em>delta7)429Ahmb&lt;/sub&gt;, Heterozygous for Smn1&lt;sub&gt;tm1Msd&lt;/sub&gt;, Homozygous for Tg(SMN2</em>delta7)429Ahmb</td>
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<td>5 weeks</td>
<td>Female</td>
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<tr>
<td>5 weeks</td>
<td>Male</td>
<td>Homozygous for Grm7&lt;sub&gt;Tg(SMN2<em>delta7)429Ahmb&lt;/sub&gt;, Heterozygous for Smn1&lt;sub&gt;tm1Msd&lt;/sub&gt;, Homozygous for Tg(SMN2</em>delta7)429Ahmb</td>
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<td>6 weeks</td>
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<tr>
<td>6 weeks</td>
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<td>7 weeks</td>
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<tr>
<td>7 weeks</td>
<td>Male</td>
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<td>8 weeks</td>
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<tr>
<td>8 weeks</td>
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<th>BREEDER PAIR</th>
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<td>Female</td>
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Email: TechTran@jax.org

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