These mice exhibit a molecular and progressive neurodegenerative phenotype similar to Type III spinal muscular atrophy.

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 murine models of Spinal Muscular Atrophy. See our full service platform.

Donating Investigator
Hung Li, Institute of Molecular Biology

GENETIC OVERVIEW

<table>
<thead>
<tr>
<th>Genetic Background</th>
<th>Generation</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>N10F30</td>
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<td></td>
<td>(2020-11-25 00:00:00)</td>
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</table>

**Smn1**<sup>tm1Hung</sup>

<table>
<thead>
<tr>
<th>Allele Type</th>
<th>Gene Symbol</th>
<th>Gene Name</th>
</tr>
</thead>
<tbody>
<tr>
<td>Targeted (Null/Knockout)</td>
<td>Smn1</td>
<td>survival motor neuron 1</td>
</tr>
</tbody>
</table>

**Tg(SMN2)2Hung**

<table>
<thead>
<tr>
<th>Allele Type</th>
<th>Gene Symbol</th>
<th>Gene Name</th>
</tr>
</thead>
<tbody>
<tr>
<td>Transgenic (Inserted expressed sequence, Humanized sequence)</td>
<td></td>
<td></td>
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</tbody>
</table>

RESEARCH APPLICATIONS
Mice that are homozygous for the $Smn1^{tm1Hung}$ knock-out allele and homozygous for the Tg(SMN2)2Hung transgene display a varied Type III SMA phenotype - they are viable, fertile and exhibit short and thickened tails. RT-PCR analysis detects alternative splicing of the transgene. Histological examination of tail tissue reveals atrophic muscles and subcutaneous edema. Skeletal muscle tissue has fewer myocytes and atrophic muscle bundles. Large motor neurons in the anterior horns of the spinal cord degenerate and are lost. There is a strong correlation between estimated copy number of the transgene and severity of the neurodegenerative phenotype. Of note, mice hemizygous for Tg(SMN2)2Hung have ~2 copies of the transgene, whereas mice homozygous for Tg(SMN2)2Hung have ~4 copies of the transgene.

Mice homozygous for $Smn1^{tm1Hung}$ and hemizygous for Tg(SMN2)2Hung display a Type I SMA-like phenotype and die at ~14 days of age.

Mice that are heterozygous for $Smn1^{tm1Hung}$ and hemizygous for Tg(SMN2)2Hung are phenotypically normal.

Non-transgenic (noncarrier) mice are phenotypically normal when heterozygous for $Smn1^{tm1Hung}$, but embryonic lethal when homozygous for $Smn1^{tm1Hung}$ - failing to survive past embryonic day 6.5.

Importation of this model was supported by the Spinal Muscular Atrophy Foundation.
Genetics

Smn1<sup>tm1Hung</sup>

Tg(SMN2)2Hung

Disease/Phenotype

Disease Terms

Research Areas By Phenotype

Mammalian Phenotype Terms by Genotype

References

Technical Support

Genotyping Protocols
Standard PCR: Smn1
Standard PCR: Smn1
Standard PCR: Tg(SMN2)2Hung
QPCR: Tg(SMN2)2Hung

Genotyping resources and troubleshooting

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

The *Smn1* (survival motor neuron 1) gene on Chr 13 and the randomly inserted transgene are not linked and will segregate independently.

Mice homozygous for the *Smn1*\textsuperscript{\textit{tm1Hung}} knock-out allele and homozygous for the Tg(SMN2)2Hung transgene (double homozygous mice) are viable and fertile with a varied Type III SMA phenotype. The Jackson Laboratory Stock No. 005058 is routinely maintained by breeding double homozygous mice together [i.e., breeding HOM HOM x HOM HOM].

Of note, breeding double homozygous mice from Stock No. 005058 to mice from the *Smn1* heterozygous non-transgenic control line (Stock No. 031678) would allow researchers to generate the following offspring:

i. homozygous for *Smn1*\textsuperscript{\textit{tm1Hung}} and hemizygous for Tg(SMN2)2Hung [i.e., HOM HEMI; ~50%] - these mice display a Type I SMA-like phenotype and die at ~14 days of age 

ii. heterozygous for *Smn1*\textsuperscript{\textit{tm1Hung}} and hemizygous for Tg(SMN2)2Hung [i.e., HET HEMI; ~50%] - these mice are phenotypically normal

Additional Breeding and Husbandry Support

Mating System

Homozygous Smn1\textsuperscript{tm1Hung}, Homozygous Tg(SMN2)2Hung x Homozygous Smn1\textsuperscript{tm1Hung}, Homozygous Tg(SMN2)2Hung

Citation

When using the SMA-like mouse strain in a publication, please cite the originating article(s) and include JAX stock #005058 in your Materials and Methods section.

Animal Health Reports

Facility Barrier Level Descriptions

- AX11 (Maximum)

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**Pricing & Availability**

Live mice available in varying quantities. Ask Customer Service for details.

**Domestic\&International**

Pricing effective for USA, Canada and Mexico shipping destinations

<table>
<thead>
<tr>
<th>AGE</th>
<th>SEX</th>
<th>GENOTYPE</th>
<th>PRICE</th>
</tr>
</thead>
<tbody>
<tr>
<td>4 weeks</td>
<td>Female</td>
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<tr>
<td></td>
<td>Male</td>
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<td>Female</td>
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<tr>
<td></td>
<td>Male</td>
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<tr>
<td>6 weeks</td>
<td>Female</td>
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<tr>
<td></td>
<td>Male</td>
<td>Homozygous for Smn1\textsuperscript{\textit{tm1Hung}}, Homozygous for Tg(SMN2)2Hung</td>
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<tr>
<td>7 weeks</td>
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<tr>
<td></td>
<td>Male</td>
<td>Homozygous for Smn1\textsuperscript{\textit{tm1Hung}}, Homozygous for Tg(SMN2)2Hung</td>
<td>$255.00</td>
</tr>
</tbody>
</table>
### RELATED PRODUCTS AND SERVICES

| Frozen Mouse Embryo | FVB.Cg-Smn1<tm1Hung> Tg(SMN2)2Hung/J Frozen Embryos | $2595.00 |

### BREEDER PAIR

<table>
<thead>
<tr>
<th>SEX</th>
<th>GENOTYPE</th>
<th>PRICE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Female</td>
<td>Homozygous for Smn1&lt;tm1Hung&gt;, Homozygous for Tg(SMN2)2Hung</td>
<td>$510.00</td>
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<tr>
<td>Male</td>
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</tbody>
</table>

### 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.

Terms Of Use
ADDITIONAL USE RESTRICTIONS APPLY

Use of MICE by non-profits requires a Material Transfer Agreement (MTA) and for-profit entities require a license.

LICENSING INFORMATION
Phone: 207-288-6470
Email: TechTran@jax.org

Related Strains