Overview

Exon 2 of the mouse Ndufs4 gene has been excised in this knockout allele, useful in studies of mitochondrial disorders.

Donating Investigator

Dr. Richard Palmiter, University of Washington
Details

Detailed Description

Mitochondrial complex I (CI)-associated defects are the most common mitochondrial disorders. Defects in the non-enzymatic, nuclear-encoded CI protein NDUFSA4 (NADH dehydrogenase (ubiquinone) Fe-S protein 4) cause a Leigh-like phenotype in humans that results in death within 3–16 months after birth.

Exon 2 of the mouse Ndufs4 gene has been excised in this knockout allele. Homozygous knockout mice are smaller than normal and begin losing body hair by postnatal day 21 (P21). Hair grows back during the next hair-growth cycle. Animals have normal locomotor activity during both day and night until approximately P30, after which they become lethargic. Knockout mice are blind and cataracts sometimes appear in one or both eyes as early as P20. After P35, some knockout mice are unable to open their eyes completely and, with variable time of onset, there is a rapid decline in acoustic startle response. Starting around P35, the homozygous knockout mice develop severe ataxia. Between P35 and P50, the knockout mice stop gaining weight, display worsened ataxia, cease grooming, and die. Homozygous knockout mice die by P55 and are infertile; heterozygotes appear normal for at least a year.
Genotyping Protocols
MELT: Ndufs4<sup>tm1.1Rpa</sup>
Genotyping resources and troubleshooting

Breeding Considerations
Homozygous mice are viable and fertile. Homozygous knockout mice die by postnatal day 55 and are infertile.

Additional Breeding and Husbandry Support

Mating System
Heterozygote x Wild-type
Wild-type x Heterozygote

Citation
If you use the Ndufs4<sup>tm1.1Rpa</sup> / J mouse strain in a publication, please cite the originating article(s) and include JAX stock #027058 in your Materials and Methods section.

Animal Health Reports
- Facility Barrier Level Descriptions
  - AX9 (Standard)
  - AX9 (Standard)

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

### Domestic
Pricing effective for USA, Canada and Mexico shipping destinations

#### Live Mouse

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<th>AGE</th>
<th>SEX</th>
<th>GENOTYPE</th>
<th>PRICE</th>
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<tr>
<td>Approx 4-8 weeks</td>
<td>Female</td>
<td>Heterozygous for Ndufs4&lt;sup&gt;Im1.1Rpa&lt;/sup&gt;</td>
<td>$255.00</td>
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<tr>
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<td>Male</td>
<td>Heterozygous for Ndufs4&lt;sup&gt;Im1.1Rpa&lt;/sup&gt;</td>
<td>$255.00</td>
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<tr>
<td>Approx 4-8 weeks</td>
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<td>Wild-type for Ndufs4&lt;sup&gt;Im1.1Rpa&lt;/sup&gt;</td>
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#### Breeder Pair

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#### Terms of Use

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Email: TechTran@jax.org
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By Allele

By Gene

By Collection