B6(Cg)-Htra2^mnd2/J

Stock No: 004608 | motor neuron degeneration 2

Spontaneous Mutation

CRYO RECOVERY

PLACE ORDER

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

Overview

Also Known As: motor neuron degeneration 2

These mice carry a spontaneous mutation at the Htra2 locus characterized by a basal ganglia disorder resulting in muscle atrophy, hunched posture, increased imbalance, chorea, dystonia, and progressive akinesis.

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GENETIC OVERVIEW
Mice homozygous for the recessive Htra2\textsuperscript{mnd2} mutation have a basal ganglia disorder initially described as an early onset motor neuron disease. This is first outwardly evident by 21 to 24 days of age as an unsteady gait with extended hind limbs, repetitive movements and episodes of sudden arrests. This progresses to include severe muscle atrophy, hunched posture, increased imbalance, chorea, dystonia, and progressive akinesis. A failure to gain weight becomes evident shortly after the onset of the other symptoms and by 35 days of age wildtype littermates are twice as heavy as the mutants. Body fat is not detectable at necropsy. Both the spleen and the thymus drop from normal weights at 23 days of age to 10% of normal at 30 days of age and the thymic corticomediulary junction is lost. Death usually occurs within two weeks of disease onset, by 40 days of age. The growth retardation is not the primary cause since disease is not delayed by intragastric feeding.

Although the initial assessment of the Htra2\textsuperscript{mnd2} phenotype described a primary motor neuron disease, it has been characterized subsequently as a basal ganglia disorder. Neurodegeneration is first detected in the striatum around 23 days of age and progresses such that mid-striatum sections at 39 days of age show a loss of approximately 50% of the neurons. Neuronal degeneration that is much less severe is also found after 30 days of age in the N. amygdaloïdes corticalis, N. subthalamicus, Globus pallidus, caustrum, amygdala, and brain stem, in addition to the motor neuron degeneration found in the spinal cord which led to the initial characterization of motor neuron degeneration. No neurodegeneration is detected in the cerebellum, and myelin staining is normal. Astrogliosis and microgliosis are detected only in the striatum and N. amygdaloïdes, and progress parallel with neurodegeneration.

There is an upregulation of Il1b, Il6, Il10, Il12, Csf1 and Tnfa in the central nervous system, an upregulation of Csf2, Il1b and Tnfa in the lungs, and an upregulation of Il1b and Tnfa in spleen as well. The dying neurons have characteristics consistent with both apoptotic and necrotic death. Transgenic expression of human BCL2 under the control of a neuron-specific enolase promoter fails to prevent disease in Htra2\textsuperscript{mnd2} homozygotes.
Histochemistry of spinal cords taken from 34-38 day old homozygotes shows that the motoneurons in both the cervical and lumbar regions are swollen, spherical, and show weaker staining with cresyl violet. Abnormal spontaneous activity with fibrillation potentials and positive waves is detected in needle electromyography of hind limb muscles, indicating denervation. However, unlike surgical and other models of denervation, Htra2<sup>md2</sup> homozygotes do not have an increase in transcription of the acetylcholine receptor alpha subunit in affected muscle. Although motor nerve conduction velocities are normal, indicating normal axon function, the compound muscle action potential amplitudes are less than half of normal.

**Development**

**Control Suggestions**

**Selected References**

**Genetics**

Htra2<sup>md2</sup>

**Disease/Phenotype**

**Disease Terms**

**Research Areas By Genotype**

**Mammalian Phenotype Terms by Genotype**

**References**

**Technical Support**

CONTACT TECHNICAL SUPPORT

**Genotyping Protocols**

Restriction Enzyme Digest: Htra2<sup>md2</sup>
Sanger sequencing: Htra2<sup>md2</sup>
Genotyping resources and troubleshooting

**Breeding Considerations**

Homozygotes die between 3-4 weeks of age and are not available for distribution.

Additional Breeding and Husbandry Support

**Citation**

If you publish the use of this mouse strain in a publication, please cite the originating article(s) and include JAX stock #004608 in your Materials and Methods section.

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

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<td>Cryo Recovery</td>
<td>Heterozygous or Wild-type for Htra2&lt;mnd2&gt;</td>
<td>$2,595.00</td>
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