B6EiC3Sn a/A-Otc $spf-ash$/J

Stock No: 001811 | sparse fur-abnormal skin and hair

Spontaneous Mutation

Also Known As: sparse fur-abnormal skin and hair

The Otc $spf-ash$ mouse model of chronic hyperammonemia is useful to study acute metabolic decompensation with hyperammonemia in response to infection/immune challenge.

**GENETIC OVERVIEW**

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<th>Genetic Background</th>
<th>Generation</th>
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<tr>
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**a**

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<tr>
<th>Allele Type</th>
<th>Gene Symbol</th>
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**Otc $spf-ash$**

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<td>Spontaneous (Hypomorph)</td>
<td>Otc</td>
<td>ornithine transcarbamylase</td>
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**RESEARCH APPLICATIONS**

Metabolism Research
Dermatology Research
Mouse/Human Gene Homologs
The urea/ureagenesis cycle functions to incorporate the normal metabolic waste product, ammonia, into urea. Ornithine transcarbamylase (OTC) is a critical enzyme in the urea cycle, and OTC-deficiency (OTCD) is the most prevalent human urea cycle disorder (UCD) characterized by life-threatening episodes of acute metabolic decompensation with hyperammonemia (HA). The sparse fur-abnormal skin and hair mutation (Otc$^{spf-ash}$) on the X chromosome is a hypomorph; resulting in only 5-10% normal hepatic OTC activity. This decreased OTC activity ultimately results in higher plasma concentration of ammonia.

OTC-deficient mice (homozygous females [Otc$^{spf-ash/spf-ash}$] and hemizygous males [Otc$^{spf-ash/Y}$]) develop hyperammonemia and present many characteristics of the human disorder. The Otc$^{spf-ash}$ mouse model of chronic hyperammonemia is useful to study acute metabolic decompensation with hyperammonemia in response to infection/immune challenge.

Specifically, compared to wildtype mice similarly treated with i.p. LPS injection, chronic hyperammonemic Otc$^{spf-ash}$ males display more pronounced and prolonged sickness behavior, as well as cognitive deficits/neurochemical changes.

Following respiratory infection with PR8 influenza (influenza A/Puerto Rico/8/34), Otc$^{spf-ash}$ mice display an altered hepatic immune response, elevated liver transaminases, increased hyperammonia and failure to increase ureagenesis during infection.

OTC-deficient mice are viable and fertile with no overt sickness in specific pathogen free conditions, but are significantly smaller than mature 6-7 month wildtype mice. Previous reports indicate that hemizygous males may need to remain with the mother until 4-6 weeks of age, and powdered food should be added to the weaning cage. Additionally, breeding heterozygous females (Otc$^{spf-ash/+}$) with hemizygous males is not very productive. Because it is difficult to reliably distinguish heterozygous females from wildtype females by phenotype, the B6EIC35nF1/J female x hemizygous male breeding scheme can be utilized to generate heterozygous females (as all female pups should be Otc$^{spf-ash/+}$).
Disease/Phenotype

Disease Terms

Research Areas By Genotype

Mammalian Phenotype Terms by Genotype

References

Technical Support

CONTACT TECHNICAL SUPPORT

Genotyping Protocols
Sanger sequencing: Otc<sup>spf-ash</sup>-SEQ
Genotyping resources and troubleshooting

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

Breeding Considerations
The Otc<sup>spf-ash</sup> allele is on the X chromosome. When maintaining a live colony at The Jackson Laboratory, B6EiC3SnF1/J females (Stock No. 001875) are bred with hemizygous males (Otc<sup>spf-ash/Y</sup>) or as heterozygous females (Otc<sup>spf-ash/Y</sup> x B6EiC3SnF1/J males (Stock No. 001875)).

OTC-deficient mice (homozygous females [Otc<sup>spf-ash/spf-ash</sup>] and hemizygous males) are viable and fertile with no overt sickness in specific pathogen free conditions, but are significantly smaller than mature 6-7 month wildtype mice. Previous reports indicate that hemizygous males may need to remain with the mother until 4-6 weeks of age, and powdered food should be added to the weaning cage. Additionally, breeding heterozygous females (Otc<sup>spf-ash/ Y</sup>) with hemizygous males is not very productive. Because it is difficult to reliably distinguish heterozygous females from wildtype females by phenotype, the B6EiC3SnF1/J female x hemizygous male breeding scheme can be utilized to generate heterozygous females (as all female pups should be Otc<sup>spf-ash/+</sup>).

Additional Breeding and Husbandry Support

Mating System
F1 x Hemizygote
B6EiC3SnF1/J (001875) x hemizygot
Heterozygote x F1
Heterozygous x B6EiC3SnF1/J (001875)

Appearance
black, small body size, sparse fur
Related Genotype: a/a Otc<sup>spf-ash/Y</sup>

agouti, small body size, sparse fur
Related Genotype: A/? Otc<sup>spf-ash/Y</sup>

black, normal fur and body size
Related Genotype: a/a +/- or a/a +/- Y

agouti, normal fur and body size
Related Genotype: A/? +/- or A/? +/- Y
### Pricing & Availability

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

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<th>AGE</th>
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Related Products and Services

Frozen Mouse Embryo $2,595.00 per straw or vial

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

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All

By Allele

By Gene

By Collection

All Related Strains
Leading the search for
TOMORROW'S CURES