B6.Cg-Gusb mps/BrkJ

Stock No: 006407 | B6.MPS VII-H2 b

Congenic Spontaneous Mutation

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

Also Known As: B6.MPS VII-H2 b

These mice carry a spontaneous intragenic deletion in the Gusb gene resulting in the failure to express the lysosomal enzyme beta glucuronidase. They exhibit various skeletal and connective tissue anomalies and are useful in studies of lysosomal storage diseases.

Donating Investigator

Brian Soper, The Jackson Laboratory

Genetic Background | Generation

| Gusb mps |

<table>
<thead>
<tr>
<th>Allele Type</th>
<th>Gene Symbol</th>
<th>Gene Name</th>
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<tr>
<td>Spontaneous</td>
<td>Gusb</td>
<td>glucuronidase, beta</td>
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View Genetics

Sensorineural Research
Developmental Biology Research
Neurobiology Research
Mouse/Human Gene Homologs
Internal/Organ Research
Metabolism Research
Research Tools

View All Research Applications
Details

Detailed Description

Mice homozygous for the "mps" (mucopolysaccharidosis type VII or MPS VII) mutation are devoid of expression of the lysosomal enzyme beta glucuronidase. Homozygous animals are viable, but females have a deficiency in lactation. Skeletal and connective tissue anomalies in both males and females are believed to prevent successful breeding. As this mutation is recessive, heterozygous mice are phenotypically similar to wildtype. Homozygotes exhibit short and thickened long bones (smaller than heterozygous or wildtype littermates), "pug type" appearance of the nose, hepatomegaly, splenomegaly, corneal clouding, and deafness. These mice have the H2b haplotype typical of inbred C57BL/6 mice. MPS VII mice are a model of the beta glucuronidase enzyme deficiency in humans called Sly Disease. They may be useful in developing new therapies (enzyme replacement, cell transplantation, gene therapy) broadly applicable to other lysosomal storage diseases.

Development

Control Suggestions

Selected References

Genetics

Gusb<sup>mps</sup>

Disease/Phenotype

Disease Terms

Research Areas By Genotype

Mammalian Phenotype Terms by Genotype

References

Technical Support
Genotyping Protocols
Restriction Enzyme Digest: Gusb<sup>mps</sup>
Pyrosequencing: Gusb<sup>mps</sup>
Sanger sequencing: Gusb<sup>mps</sup>-SEQ
Genotyping resources and troubleshooting

Breeding Considerations
When maintaining a live colony, heterozygous mice are bred together. Mice homozygous for this recessive mutation are viable, but exhibit breeding problems.
Additional Breeding and Husbandry Support

Citation
When using the B6NRS VII Hp<sup>b</sup> mouse strain in a publication, please cite the originating article(s) and include JAX stock #006407 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
Typically mice are recovered in 10-14 weeks. Contact Customer Service to place an order or for more information.

<table>
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<tr>
<th>SERVICE</th>
<th>GENOTYPE</th>
<th>PRICE</th>
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<tbody>
<tr>
<td>Cryo Recovery</td>
<td>Heterozygous for Gusb&lt;sup&gt;mps&lt;/sup&gt;</td>
<td>$2,595.00</td>
</tr>
</tbody>
</table>

We will fulfill your order by providing at least two carriers for each strain ordered. The total number, sex, and genotypes provided will vary, although typically 8 or more animals are provided. Please check genotypes which will be recovered. While the genotypes of all animals produced will be communicated to you prior to scheduling shipment, the genotypes of animals provided may not reflect the mating scheme and genotypes described in the strain description. Animals are typically ready to ship in 11-14 weeks. If a second recovery is required to produce the minimum number of animals, then delivery time would increase to approximately 25 weeks. If we fail to produce animals of the correct genotype, you will not be charged. We cannot guarantee the reproductive success of mice shipped to your facility. If the mice are lost after the first three days (post-arrival) or do not produce progeny at your facility, a new order and fee will be necessary.

Cryorecovery to establish a Dedicated Supply for greater quantities of mice. Mice recovered can be used to establish a dedicated colony to contractually supply you mice according to your requirements. Price by quotation.

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