

C3H/HeO_uJ-*Gusb*^{mps-2J}/BrkJ

Stock No: **003525** | mucopolysaccharidosis VII 2 Jackson

 Spontaneous Mutation

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storage disease and elevated levels of the lysosomal enzymes alpha-galactosidase and beta-hexosaminidase.

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GENETIC OVERVIEW

Genetic Background Generation

Gusb^{mps-2J}

Alele Type	Gene Symbol	Gene Name
Spontaneous	<i>Gusb</i>	glucuronidase, beta

VIEW GENETICS

RESEARCH APPLICATIONS

Developmental Biology Research
Metabolism Research
Mouse/Human Gene Homologs

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Details

Detailed Description

Mice homozygous for the Gus^{mps-2J} allele exhibit a phenotype similar to Gus^{mps} homozygotes including skeletal deformities, lysosomal storage disease and elevated levels of the lysosomal enzymes alpha-galactosidase and beta-hexosaminidase (Gwynn et al., 1998). Like the Gus^{mps} heterozygote, Gus^{mps-2J} heterozygotes have a 26-85% reduction in beta-glucuronidase activity, depending on tissue type (Gwynn et al., 1998, Birkenmeier et al., 1989). Homozygotes of both alleles have beta-glucuronidase activity levels at 1% of the control level. Unlike the Gus^{mps} homozygote, Gus^{mps-2J} homozygotes live longer, are fertile and can raise litters to weaning age (Gwynn et al., 1998). In addition to a difference in the nature of the mutations between these two alleles, it is also likely that the phenotypic differences are the result of strain background. C3H/HeOuj mice carry a different set of alleles at the *Gus* complex than C57BL/6J mice. Beta-glucuronidase activity in C3H/HeOuj mice is 10-38% that of C57BL/6J mice (Gwynn et al., 1998). The *Gus* complex alleles determine the rate of enzyme synthesis and may modulate the expression of the beta-glucuronidase deficiency. This strain is a model for the human lysosomal storage disease, mucopolysaccharidosis type VII.

Development

Control Suggestions

Selected References

Genetics

$Gusb^{mps-2J}$

Disease/Phenotype

Disease Terms

Research Areas By Phenotype

[+ Mammalian Phenotype Terms by Genotype](#)

[+ References](#)

[- Technical Support](#)

C O N T A C T T E C H N I C A L S U P P O R T

Genotyping Protocols

Standard PCR:[Gusb](#)

[Genotyping resources and troubleshooting](#)

Appearance

black

Related Genotype: *a/a*

Citation

When using the mucopolysaccharidosis VII 2 Jackson mouse strain in a publication, please [cite the originating article\(s\)](#) and include JAX stock #003525 in your Materials and Methods section.

Animal Health Reports

[Facility Barrier Level Descriptions](#)

Production of mice from cryopreserved embryos or sperm occurs in a maximum barrier room, [G200](#)

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SERVICE/PRODUCT	DESCRIPTION	PRICE
Cryo Recovery	Heterozygous for usb<mps-2J>	\$2,854.50

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C3H/HeO_uJ-Gusb<mps-2J>/BrkJ

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