Overview

Homozygous mice for this targeted mutation in beta glucuronidase (Gusb) exhibit growth retardation, shortened extremities, and facial dysmorphism. This model has increased urinary glycosaminoglycan and secondary elevation of other lysosomal storage enzymes. This strain represents a model of human GUS deficiency characterized by the most prevalent human mutation, L176F, and is associated with mild Mucopolysaccharidosis type VII (MPS VII or Sly Syndrome).

Donating Investigator

William S Sly, Saint Louis University Medical Center
### GENETIC OVERVIEW

**Genetic Background**

**Generation**

<table>
<thead>
<tr>
<th>Allele Type</th>
<th>Gene Symbol</th>
<th>Gene Name</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gusb&lt;sup&gt;tm3Sly&lt;/sup&gt;</td>
<td>Gusb</td>
<td>glucuronidase, beta</td>
</tr>
</tbody>
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### RESEARCH APPLICATIONS

- Developmental Biology Research
- Metabolism Research
- Internal/Organ Research

### BASE PRICE

**Starting at:**

$2,854.50 Domestic price Cryo Recovery

### Details

Homozygous mice are viable but infertile. Fertility can be restored with enzyme replacement therapy. Less than 1% residual enzyme activity is observed. Homozygous mice are indistinguishable from wild type mice at birth, but show distinct growth retardation, shortened extremities, and facial dysmorphism observable at three months with a moderate increase in severity with age. Long bones of the lower extremities are shortened, broad, and sclerotic. Compared to wild-type, this model has increased urinary glycosaminoglycan and secondary elevation of other lysosomal storage enzymes. Further, homozygous mice show abundant lysosomal storage in liver, kidney, leptomeningeal cells, cornea, and retinal pigment epithelium. The phenotype exhibited by this mutant mouse strain is less severe than that observed in a similarly constructed mutant (Stock No. 005643) which bears a E536A point mutation. This strain represents a model of human GUS deficiency characterized by the most prevalent human mutation, L176F, and is associated with mild Mucopolysaccharidosis type VII (MPS VII or Sly Syndrome).

### Development
Genotyping Protocols
Pyrosequencing: \textit{Gusb}^{tm3Sly}
Genotyping resources and troubleshooting

Breeding Considerations
When maintaining a live colony, heterozygotes are intercrossed to produce homozygotes, heterozygotes, and wild type mice. Homozygous mice are infertile. Intravenous injection of beta-glucuronidase into young homozygous males will restore fertility (see Sands et al, 1994 J Clin Inv 93:2324-2331).

Additional Breeding and Husbandry Support

Citation
When using the B6.129X-\textit{Gusb}^{tm3Sly}/J mouse strain in a publication, please cite the originating article(s) and include JAX stock #005644 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

Pricing & Availability

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

Payment Terms and Conditions
Terms are granted by individual review and stated on the customer invoice(s) and account statement. These transactions are payable in U.S. currency within the granted terms. Payment for services, products, shipping containers, and shipping costs that are rendered are expected within the payment terms indicated on the invoice or stated by contract. Invoices and account balances in arrears of stated terms may result in The Jackson Laboratory pursuing collection activities including but not limited to outside agencies and court filings.

The Jackson Laboratory's Genotype Promise
The Jackson Laboratory has rigorous genetic quality control and mutant gene genotyping programs to ensure the genetic background of JAX® Mice strains as well as the genotypes of strains with identified molecular mutations. JAX® Mice strains are only made available to researchers after meeting our standards. However, the phenotype of each strain may not be fully characterized and/or captured in the strain data sheets. Therefore, we cannot guarantee a strain's phenotype will meet all expectations. To ensure that JAX® Mice will meet the needs of individual research projects or when requesting a strain that is new to your research, we suggest ordering and performing tests on a small number of mice to determine suitability for your particular project.
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All

By Allele

By Gene