These mice carry the mouse mucopolysaccharidosis VII mutation on a C57BL/6J with a dominant spotting genetic background and exhibit lysosomal storage disease. They may serve as a model for Sly disease in humans.
GENETIC OVERVIEW

<table>
<thead>
<tr>
<th>Genetic Background</th>
<th>Generation</th>
</tr>
</thead>
</table>

**Gusb**

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

**Kit**

<table>
<thead>
<tr>
<th>Allele Type</th>
<th>Gene Symbol</th>
<th>Gene Name</th>
</tr>
</thead>
<tbody>
<tr>
<td>Spontaneous</td>
<td>Kit</td>
<td>KIT proto-oncogene receptor tyrosine kinase</td>
</tr>
</tbody>
</table>

RESEARCH APPLICATIONS

Cancer Research  
Mouse/Human Gene Homologs  
Research Tools  
Neurobiology Research  
Sensorineural Research  
Reproductive Biology Research  
Dermatology Research  
Developmental Biology Research

BASE PRICE

Starting at: $2,854.50 Domestic price Cryo Recovery
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. In appearance, homozygous Kit<sup>W-41J</sup> mice are mostly white with black eyes and brown or grey spots. They are fertile, distinct from mice with other alleles of Kit, and have the impaired hemopoiesis causing mild, normochromic, macrocytic anemia. 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. This strain, with combined Kit<sup>W-41J</sup> and Gusb<sup/mps</sup>, provides a genetically myeloablated population in which to study stem cell engraftment and reconstitution.
Breeding Considerations
When maintaining a live colony, these mice can be bred as heterozygotes. Homozygous \( Gusb^{mps} \) females do not lactate. Homozygous \( Gusb^{mps} \) males do not breed.

Additional Breeding and Husbandry Support

Citation
When using the WMPS VII mouse strain in a publication, please cite the originating article(s) and include JAX stock #006564 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>
<thead>
<tr>
<th>SERVICE/PRODUCT</th>
<th>DESCRIPTION</th>
<th>PRICE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cryo Recovery</td>
<td>Homozygous for Kit&lt;W-41J&gt;, Heterozygous or Wild-type for Gusb&lt;mps&gt;</td>
<td>$2,854.50</td>
</tr>
</tbody>
</table>

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.

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Email: TechTran@jax.org
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## Related Strains

- **All**
- **By Allele**
- **By Gene**
- **By Collection**