Also Known As: woozy

These mice carry a spontaneous mutation at the **Sil1** locus characterized by adult-onset ataxia with cerebellar Purkinje cell loss. They may be useful in studies of Alzheimer and Parkinson diseases.
Homozygous woozy (Sil\textsuperscript{1\textasciitilde}) mice develop adult-onset ataxia with cerebellar Purkinje cell loss. Affected cells have intracellular protein accumulations reminiscent of protein inclusions in both the ER and the nucleus. In addition, upregulation of the unfolded protein response, suggestive of ER stress, occurs in mutant Purkinje cells. The wz mutation disrupts the gene Sil1 that encodes an adenine nucleotide exchange factor of BiP, a crucial ER chaperone. Thus, perturbation of ER chaperone function in terminally differentiated neurons leads to protein accumulation, ER stress and subsequent neurodegeneration.
Genotyping Protocols

Standard PCR: Sil1
Genotyping resources and troubleshooting

Citation

When using the woozy mouse strain in a publication, please cite the originating article(s) and include JAX stock #003777 in your Materials and Methods section.

Animal Health Reports

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.

Cryo Recovery

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<thead>
<tr>
<th>SERVICE/PRODUCT</th>
<th>DESCRIPTION</th>
<th>PRICE</th>
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<tr>
<td>Cryo Recovery</td>
<td>Heterozygous for Sil1&lt;wz&gt;</td>
<td>$2,854.50</td>
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