CBACa.129S6(Cg)-Ush1c<sup>tm18kts</sup>/J

Stock No: 012426

pritecic TARGETED MUTATION

CRYO RECOVERY

PLACE ORDER

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

These Ush1c216A mice contain a mutation designed to replace gene function of endogenous mouse Ush1c with the human USH1C. They also contain a mutation (216G->A) designed to retain the endogenous mouse sequence. These humanized mice develop both deafness and retinal degeneration, as evident by absent auditory-evoked brainstem responses and progressive loss of rod photoreceptors, providing a novel resource to study the disease mechanism and the development of therapies.

**Donating Investigator**

Bruce Temple, University of Washington

<table>
<thead>
<tr>
<th>Genetic Background</th>
<th>Generation</th>
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<tbody>
<tr>
<td><strong>Ush1c&lt;sup&gt;tm18kts&lt;/sup&gt;</strong></td>
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<thead>
<tr>
<th>Allele Type</th>
<th>Gene Symbol</th>
<th>Gene Name</th>
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<tbody>
<tr>
<td>Targeted (Humanized sequence)</td>
<td>Ush1c</td>
<td>USH1 protein network component harmonin</td>
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**Mouse/Human Gene Homologs**

Sensorineural Research
Developmental Biology Research
Neurobiology Research

**VIEW GENETICS**

**VIEW ALL RESEARCH APPLICATIONS**
Details

Detailed Description

These mice contain a mutation designed to replace gene function of the endogenous mouse Ush1c gene with the human USH1C function. They also contain a mutation (216G→A) designed to retain the endogenous mouse Ush1c sequence. Mice with this mutation carry the same truncated USH1C mRNA transcript and splicing defect as found in the cochleae and retinas of Usher 1C patients. Mice homozygous for Ush1c<sup>216A</sup> are viable, fertile, and exhibit behavioral characteristics of deaf mice; they are hyperactive, display circling and head tossing behavior, and do not have a Preyer reflex. They also develop retinal degeneration, as evident by absent auditory-evoked brainstem responses and progressive loss of rod photoreceptors, providing a novel resource to study the disease mechanism and the development of therapies.

Development

Expression Data

Control Suggestions

Selected References

Genetics

Ush1c<sup>tm1</sup>lks

Disease/Phenotype

Disease Terms

Research Areas By Genotype

Mammalian Phenotype Terms by Genotype

References

Technical Support
Genotyping Protocols
Genotyping resources and troubleshooting

Breeding Considerations
When maintaining a live colony, homozygous mice may be bred together.
Additional Breeding and Husbandry Support

Citation
When using the PRAGS 129S1/CD1-Ush1c<tm1Bkts>/J mouse strain in a publication, please cite the originating article(s) and include JAX stock #012426 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.

Domestic | International
Pricing effective for USA, Canada and Mexico shipping destinations

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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 or wildtype for Ush1c&lt;tm1Bkts&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.

Related Products and Services

<table>
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<tr>
<th>SERVICE</th>
<th>PRICE</th>
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<tbody>
<tr>
<td>Frozen Mouse Embryo</td>
<td>$2,595.00 per straw or vial</td>
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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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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.

### Terms Of Use

**General Terms and Conditions**

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Phone: 207-288-6470  
Email: TechTran@jax.org

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Related Strains

All

By Allele

By Gene

By Collection

All Related Strains

ELLSWORTH FACILITY OFFICIALLY OPEN AND OPERATIONAL
Shipments have begun at our new facility, allowing increased access to JAX mouse models.

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TOMORROW'S CURES