B6.Cg-Foxp3 #/J

Stock No: 004088 | B6-scurfy

Congenic

PLACE ORDER

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

Overview

Also Known As: B6-scurfy

Scurfy mice have defective T cell tolerance leading to an X-linked lymphoproliferative disease that parallels the X-linked autoimmunity-allergic disregulation syndrome (XLAAD) in humans. Hemiogyous males are characterized by running, scaly, crusty skin on the eyelids, ears and tails, dermal thickening, squinting eyes, cachexia, reddening and swelling of the genital papilla, and small testicles that are retained in the abdominal cavity. Homozygous scurfy females develop the same disease phenotype seen in hemizygous males, but they have a normal reproductive tract.

GENETIC OVERVIEW

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<th>Genetic Background</th>
<th>Generation</th>
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**RESEARCH APPLICATIONS**
Dermatology Research
Developmental Biology Research
Endocrine Deficiency Research
Hematological Research
Mouse/Human Gene Homologs
Immunology, Inflammation and Autoimmunity Research
Cell Biology Research
Internal/Organ Research

**BASE PRICE**
Starting at:

$2,854.50 Domestic price Cryo Recovery

**Details**

**Detailed Description**

Scurvy mice develop an X-linked lymphoproliferative disease resulting from defective T cell tolerance. Phenotypes associated with these mice include running, scaly, crusty skin on the eyelids, ears and tails, dermal thickening, squinted eyes, cachexia, reddening and swelling of the genital papilla, and small testicles that are retained in the abdominal cavity. This disorder, which parallels X-linked autoimmunity-allergic regulation syndrome (XLAAD) in humans, results in Coombs' test-positive anemia, hypergammaglobulinemia, a small, thin thymus, and lymphohistiocytic proliferation in the skin and lymphoid organs, with splenomegaly, lymphadenomegaly, and hepatomegaly. Foxp3<sup>sf</sup>/Y males generally die by 16-25 days of age. Transgenic expression of Foxp3 prevents scurvy disease in Foxp3<sup>sf</sup>/Y mice.

Neonatal thymectomy of scurvy males ameliorates disease and increases lifespan; athymic nude (Foxn<sup>1</sup> nu/Foxn<sup>1</sup> nu) Foxp3<sup>sf</sup>/Y mice do not develop scurvy. While Cd4<sup>+</sup> peripheral T cells from scurvy mice can transfer the scurvy disease phenotype to wild type, histocompatible Foxn<sup>1</sup> nu/Foxn<sup>1</sup> nu or Prkdc<sup>scid</sup>/Prkdc<sup>scid</sup> hosts, bone marrow transplantation from scurvy homozygotes fails to transfer disease. Also, neither neonatal inoculation with wild type bone marrow, nor thymic lobe transplants from wild type donors into carrier males prevents disease. Northern blot analysis of skin, lymph nodes and spleen revealed over-expression of Il2, Il4, Il5, Il10, Il6, Ifng, and Tnfα; over-expression of these last three is especially high. Peripheral Cd4<sup>+</sup> T cells from scurvy mice are hyper-responsive to antigen, have an activated phenotype (Cd44<sup>+</sup>, Cd69<sup>+</sup>, Cd25<sup>+</sup>, Cd80<sup>+</sup>, Cd86<sup>+</sup>), a decreased requirement for Cd28 co-stimulation, and a decreased sensitivity to tyrosine kinase inhibitors and cyclosporin A. Prenatal or neonatal injection with anti-Cd4
antibodies can delay the onset of disease, as can the targeted disruption of Cd4. Cd8⁺ cells do not transfer disease, and targeted disruption of B2m does not alter disease onset. Activation of peripheral T cells is necessary to initiate the scurfy pathology; Foxp3sf/Y mice carrying a transgene for an ovalbumin-specific TCR and a targeted mutation of Rag1 fail to develop the scurfy disease phenotype until challenged with ovalbumin. Foxp3st homozygous females can not be generated through traditional breeding because carrier males die by 25 days of age. By breeding nude Foxp3sf/Y males with Foxp3sf/+ females, however, homozygous scurfy females can be generated that are heterozygous for the recessive Foxn1nu mutation. These homozygous scurfy females develop the same disease phenotype seen in hemizygous males, but they have a normal reproductive tract.
Production of mice from cryopreserved embryos or sperm occurs in a maximum barrier room, G200

# Pricing & Availability

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<th>SERVICE</th>
<th>GENOTYPE</th>
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<tr>
<td>Cryo Recovery</td>
<td>Heterozygous females and Wildtype males for F0xp3&lt;sf&gt;</td>
<td>$2,854.50</td>
</tr>
</tbody>
</table>

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.

## Related Products and Services

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<th>Price</th>
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<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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Terms of Use

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Email: TechTran@jax.org

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