Overview

This Ednrβ knock-out exhibits an aganglionic megacolon, skin pigmentation deficiency and lymphoid depletion. They may be used as a model for human Hirschsprung's disease.
Exon 3 is replaced by a neomycin resistance cassette in this knock-out mutant of the endothelin receptor type B (Ednrb) gene. Ednrb encodes a G protein-coupled receptor expressed in vascular endothelial cells where it is involved in vasoconstriction, vasodilation, bronchoconstriction and cell proliferation. In humans, mutations in this gene have been associated with Waardenburg syndrome type 4 (Waardenburg-Shah syndrome) which is characterized by deafness, neural crest defects, pigmentation changes, and Hirschsprung’s disease. Hirschsprung’s disease is primarily an intestinal disorder in which colon segments of variable length lack ganglion cells resulting in intestinal blockage and megacolon. Homozygous knock-out mice are initially viable but die within the first month. They show a disruption of neural crest lineage development, which is characterized by a lack of hair or skin pigmentation on
90% of their body. Death occurs by sepsis, likely from enterocolitis. Immunosuppression is evidenced by small spleens, bone marrow suppression, and thymic involution caused by the aganglionic megacolon. The phenotype is more severe (earlier onset enterocolitis and shorter life-span) on a congenic C57BL/6J strain than on the B6/129 hybrid background (Stock No. 003295). They also do not breed as well and have smaller litters than the B6/129 line. This mutation is allelic with the piebald lethal spontaneous mutation.

Development

Control Suggestions

Selected References

Genetics

Ednrb<sup>tm1Ywa</sup>

Disease/Phenotype

Disease Terms

Research Areas By Genotype

Mammalian Phenotype Terms by Genotype

References

Technical Support

Genotyping Protocols

Standard PCR: Ednrb<sup>tm1Ywa</sup>

Genotyping resources and troubleshooting

Breeding Considerations

When maintaining a live colony, heterozygous mice may be bred to wildtype mice from the colony or to C57BL/6J inbred mice (Stock No. 000664). Homozygotes are viable but usually die within the first month of life.

Additional Breeding and Husbandry Support

Citation

When using the B6.129S7-Ednrb<sup>tm1Ywa</sup>/FrykJ mouse strain in a publication, please cite the originating article(s) and include JAX stock #021933 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

Cryo Recovery

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

<table>
<thead>
<tr>
<th>SERVICE</th>
<th>GENOTYPE</th>
<th>PRICE</th>
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</thead>
<tbody>
<tr>
<td>Cryo Recovery</td>
<td>Heterozygous or wildtype for Ednrb&lt;tm1Ywa&gt;</td>
<td>$2,854.50</td>
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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

| Frozen Mouse Embryo | B6.129S7-Ednrb<tm1Ywa>/FrykJ | $2,595.00   |

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

General Terms and Conditions
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