B6(Cg)-Gt(ROSA)26Sor Im1.1(DUX4)Fij/J

Stock No: 028710 | FLEXDUX4

Targeted Mutation

AVAILABLE

PLACE ORDER

Live mice available in varying quantities. Ask Customer Service for details.
Also Known As: Dux4-fl, FLEXDUX4

FLEXDUX4 mice conditionally express human DUX4 following Cre-mediated recombination. This strain may be useful for studying facioscapulohumeral muscular dystrophy (FSHD).

Donating Investigator

Peter L. Jones, University of Nevada School of Medicine

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**GENETIC OVERVIEW**

<table>
<thead>
<tr>
<th>Genetic Background</th>
<th>Generation</th>
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<tbody>
<tr>
<td>?+pN1F7</td>
<td>(2018-08-08 00:00:00)</td>
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</tbody>
</table>

**Gt(ROSA)26Sor<sup>lm1.1(DUX4<sup>+</sup>)Plj**

<table>
<thead>
<tr>
<th>Allele Type</th>
<th>Gene Symbol</th>
<th>Gene Name</th>
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<tbody>
<tr>
<td>Targeted (Conditional ready (e.g. floxed), inserted expressed sequence, Humanized sequence)</td>
<td>Gt(ROSA)26Sor</td>
<td>gene trap ROSA 26, Philippe Soriano</td>
</tr>
</tbody>
</table>

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**RESEARCH APPLICATIONS**

Neurobiology Research
Research Tools

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**BASE PRICE**

Starting at:

$270.00 Domestic price for female 4-week

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**Details**

FLEXDUX4 mice were created using a cre-dependent one-way genetic switch (FLEX) system. Homozygotes mice carrying this DUX4 conditional allele are viable and fertile. Two sets of incompatible outward facing recombination sites (loxP and lox511) flank an
inverted human double homeobox 4 (DUX4) sequence, including exons 1-3 and both introns. The DU4X gene encodes several alternative mRNA splicing variants. The hereditary muscle disorder, facioscapulohumeral muscular dystrophy (FSHD) is caused by the toxic gain-of-function of the DU4X-full-length (DU4X-fl) mRNA isoform. The DU4X-fl mRNA, which encodes a paired homeobox domain transcription factor, is typically not expressed in healthy muscle. However, in FSHD, the rare expression of DU4X-fl (in less than 1% of muscle fibers) initiates a pathogenic cascade of events including apoptosis, differentiation defects, muscle atrophy, and susceptibility to oxidative stress. Overall, FSHD is characterized by a slowly progressing muscular dystrophy that predominantly affects the skeletal muscles of the face, scapula, and upper arms but can affect muscles of the abdomen, hip girdle, and lower legs with ~20% of patients ultimately losing ambulation.

The DU4X promoter drives expression of two short non-pathogenic isoforms (DU4X-s) and a longer cytotoxic isoform (DU4X-fl). This strain contains 4 point mutation in the 5’ splicing donor sites for the two DU4X-s mRNAs, abolishing expression of the short isoforms and only generating the pathogenic DU4X-fl mRNA isoform.

Because this construct was targeted for insertion into the Gt(Rosa)26Sor locus, DU4X-fl expression is determined by which tissue(s) express Cre recombinase. When bred to mice that express Cre recombinase, the resulting offspring will have the loxP or lox511 sites recombined, resulting in the inversion of the human DU4X-fl sequence, ending in a sense orientation.

Hemizygous and homozygous mice have low level DU4X-fl expression in the absence of Cre Recombinase. These mice exhibit alopecia, soft stool, inflammation, and muscle weakness. Homozygous are more affected, as are males compared to females.

When bred to Tg[ACTA1-cre/Esr1+12Kesr/J] mice (Stock No. 025750), resulting offspring express slightly more DU4X-fl than the FLexDU4X hemizygous mice prior to tamoxifen induction (TMX). One TMX injection of 5 mg/kg leads to a useful mildly progressive FSHD model, while 2 injections of 10mg/kg leads to a more severe model.

Of note, the donating investigator reports that when bred to Sox2-Cre mice (Stock No. 008454), resulting offspring are embryonic lethal. When bred to ACTA1-rTA, tetO-cre mice (Stock No. 012433), resulting offspring are embryonic lethal. When bred to ACTA1-Cre mice (Stock No. 006149), resulting offspring are still-born. When bred to UBC-CreERT2 mice (Stock No. 008085), without tamoxifen induction, offspring begin to show a severe neurological phenotype around 5 weeks of age, perhaps due to leaky DU4X expression in the brain.
Genotyping Protocols
High Resolution Melting: Gt(Rosa)26Sor^tm1.1(DUX4*)Pij
Genotyping resources and troubleshooting
Dietary Information
New Diet as of March 2015: Lab Diet® 5K0Q (6% fat)
Breeding Considerations
When maintaining a live colony, homozygous mice may be bred together.
Additional Breeding and Husbandry Support
Mating System
Homozygote x Homozygote

Citation
When using the Gt(Rosa)26Sor^tm1.1(DUX4*)Pij mouse strain in a publication, please cite the originating article(s) and include JAX stock #028710 in your Materials and Methods section.

Animal Health Reports
Facility Barrier Level Descriptions
AX10 (Standard)

Pricing & Availability
Live mice available in varying quantities. Ask Customer Service for details.

Live Mouse

<table>
<thead>
<tr>
<th>AGE</th>
<th>SEX</th>
<th>GENOTYPE</th>
<th>PRICE</th>
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<tbody>
<tr>
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<td>Female</td>
<td>Homozygous for Gt(Rosa)26Sor^tm1.1(DUX4*)Pij</td>
<td>$270.00</td>
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<tr>
<td></td>
<td>Male</td>
<td>Homozygous for Gt(Rosa)26Sor^tm1.1(DUX4*)Pij</td>
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<tr>
<td>5 weeks</td>
<td>Female</td>
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<tr>
<td></td>
<td>Male</td>
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<tr>
<td>6 weeks</td>
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<td></td>
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<tr>
<td>7 weeks</td>
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<tr>
<td></td>
<td>Male</td>
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<td>$270.00</td>
</tr>
<tr>
<td>Week</td>
<td>Sex</td>
<td>Genotype</td>
<td>Price</td>
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<tr>
<td>--------</td>
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<td>--------------------------</td>
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<tr>
<td>9 weeks</td>
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<tr>
<td>10 weeks</td>
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<tr>
<td>11 weeks</td>
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<tr>
<td>12 weeks</td>
<td>Female</td>
<td>Homozygous for Gt(Rosa)26 Sor^{tm1.1(DUX4)^{Pki}}</td>
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<tr>
<td></td>
<td>Male</td>
<td>Homozygous for Gt(Rosa)26 Sor^{tm1.1(DUX4)^{Pki}}</td>
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**Related Products and Services**

<table>
<thead>
<tr>
<th>Product</th>
<th>Price</th>
</tr>
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<tbody>
<tr>
<td>Frozen Mouse Embryo</td>
<td>$2,595.00 per straw or vial</td>
</tr>
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Terms of Use

General Terms and Conditions

**QUESTIONS ABOUT TERMS OF USE**

**licensing Information**

Phone: 207-288-6470

Email: TechTran@jax.org

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