

B6.Cg-Ighmbp2^{nmd-2J} Tg(Ttn-Ighmbp2)108Cx/Cx **FACULTY**

STRAIN

Stock No: **006514**

 Congenic, Targeted Mutation, Transgenic

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cardiac myopathy despite the presence of neuromuscular degeneration. This strain is useful for studies involving the role of *Ighmbp2* in motor neuron disease and human cardiomyopathy.

Donating Investigator

Gregory Cox, The Jackson Laboratory

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GENETIC OVERVIEW

Genetic Background

Generation

N10+F21
(2017-10-18 00:00:00)

Ighmbp2^{nmd-2J}

Alele Type

Spontaneous

Gene Symbol

Ighmbp2

Gene Name

immunoglobulin mu binding protein 2

Tg(Ttn-Ighmbp2)108Cx

Alele Type

Transgenic (Inserted expressed sequence)

VIEW GENETICS

RESEARCH APPLICATIONS

Neurobiology Research

Mouse/Human Gene Homologs

BASE PRICE

Starting at:

\$179.88 Domestic price for female

470.89 Domestic price for breeder pair

[VIEW PRICE LIST](#)

Details

Detailed Description

Immunoglobulin mu binding protein 2 (IGHMBP2) is a DNA/RNA helicase implicated in transcriptional regulation and mRNA splicing. Mutations in IGHMBP2 are associated with the degeneration of alpha motor neurons and spinal muscular atrophy (DSMA1, SMARD1, HMN6) as well as dilated cardiac myopathy (DCM) and skeletal myopathy. *nmd-2J* mutant mice are characterized by a progressive paralysis that begins in the hindlimbs and moves to the forelimbs in the later stages of the disease. Homozygotes initially are identified by contracted hindlimbs and impaired movement beginning at two weeks of age. Mice rarely survive past 4 weeks of age. Combining *nmd-2J* mice with transgenic mice expressing *Ighmbp2* under the control of the rat neuron-specific enolase promoter (Stock No. [003834](#), TgNI) halts neuromuscular degeneration, but reveals the presence of cardiac myopathy followed by premature death as a result of congestive heart failure (CHF).

In this strain, *nmd-2J* mice are combined with transgenic mice expressing *Ighmbp2* under the control of the muscle-specific titin (*Ttn*) promoter (TgMI), the transgene extends lifespan by preventing primary DCM despite the presence of neuromuscular degeneration. Transgene founder lines 45 and 108 exhibit similar phenotypes. This strain is useful for studies involving the role of *Ighmpb2* in motor neuron disease and human cardiomyopathy.

Development

Expression Data

Selected References

Genetics

[+ *Ighmbp2^{nmd-2J}*](#)

[+ Tg\(Ttn-Ighmbp2\)108Cx](#)

[- Disease/Phenotype](#)

[+ Disease Terms](#)

[+ Research Areas By Phenotype](#)

[+ Mammalian Phenotype Terms by Genotype](#)

[+ References](#)

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C O N T A C T T E C H N I C A L S U P P O R T

Genotyping Protocols

[Genotyping resources and troubleshooting](#)

Mating System

Tg/0 *nmd-2J*/+ x +/+ *nmd-2J*/

Citation

When using the B6.Cg-*Ighmbp2^{nmd-2J}* Tg(Ttn-Ighmbp2)108Cx/Cx mouse strain in a publication, please [cite the originating article\(s\)](#) and include JAX stock #006514 in your Materials and Methods section.

Animal Health Reports

[Facility Barrier Level Descriptions](#)

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Availability
Varies

Domestic | International

Pricing effective for USA, Canada and Mexico shipping destinations

LIVE MOUSE

AGE	SEX	GENOTYPE	PRICE
Approx 4-8 weeks	Female	Heterozygous for <i>Ighmbp2^{nmd-2J}</i> , Noncarrier	\$179.88
	Male	Heterozygous for <i>Ighmbp2^{nmd-2J}</i> , Noncarrier	\$179.88
Approx 4-8 weeks	Female	Heterozygous for <i>Ighmbp2^{nmd-2J}</i> , Hemizygous for <i>Tg(Tn-Ighmbp2)108Cx</i>	\$291.01
	Male	Heterozygous for <i>Ighmbp2^{nmd-2J}</i> , Hemizygous for <i>Tg(Tn-Ighmbp2)108Cx</i>	\$291.01

BREEDER PAIR

SEX	GENOTYPE	PRICE
Female	Heterozygous for <i>Ighmbp2^{nmd-2J}</i> , Noncarrier	\$470.89
Male	Heterozygous for <i>Ighmbp2^{nmd-2J}</i> , Hemizygous for <i>Tg(Tn-Ighmbp2)108Cx</i>	
Female	Heterozygous for <i>Ighmbp2^{nmd-2J}</i> , Hemizygous for <i>Tg(Tn-Ighmbp2)108Cx</i>	\$470.89
Male	Heterozygous for <i>Ighmbp2^{nmd-2J}</i> , Noncarrier	

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