The Jackson Laboratory

B6;CBA-Tg(ATXN3*)84.2Cce/IbezJ

Stock No: **012705**

Congenic, Transgenic

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

PLACE ORDER

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These MJD84.2 transgenic mice harbor a YAC transgene that expresses a human (*ATXN3* gene modified with an expanded 84 CAG repeat motif that is associated with Machado-Joseph disease in humans.

Donating Investigator

Ilya Bezprozvanny, UT Southwestern Medical Center at Dallas

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GENETIC OVERVIEW

Genetic Background

Generation

?+pN1
(2020-07-31 00:00:00)

Tg(ATXN3*)84.2Cce

Allele Type

Transgenic (Inserted expressed sequence, Humanized sequence)

VIEW GENETICS

RESEARCH APPLICATIONS

Developmental Biology Research
Neurobiology Research

VIEW ALL RESEARCH APPLICATIO

BASE PRICE

Starting at:

$255.00 Domestic price for female 4-week

333.51 Domestic price for breeder pair

$2,854.50 Domestic price Cryo Recovery

VIEW PRICE LIST

Details

Detailed Description

MJD84.2 transgenic mice (also called SCA3-YAC-84Q) harbor a YAC transgene that expresses a human ataxin 3 gene (*ATXN3*; also called Machado-Joseph disease [MJD], MJD1, or spinocerebellar ataxia 3 [SCA3]) modified with a human MJD/SCA3-associated 84 CAG repeat expansion [$(CAG)_2$ CAAAAGCAGCAA(CAG)78 repeat motif (Q_3KQ_{80}), also referred to as $(CAG)_{84}$]. See below for comment on CAG repeat stability. These MJD84.2 transgenic mice may be useful in studying the progressive neurodegenerative processes underlying Machado-Joseph disease/spinocerebellar ataxia 3 (MJD/SCA3) pathogenesis and other polyglutamine and trinucleotide repeat disorders.

Hemizygous mice (MJD84.2) harbor two copies of the transgene at a single genomic integration site, with transgene expression levels and patterns almost identical to endogenous MJD. Transgene expression is widespread - detected in the cerebellum, cerebral cortex, heart, lung, spleen, liver and skeletal muscle. Stable transmission of the MJD1/CAG84 transgene has been demonstrated for multiple generations with a predicted frequency of about 50%.

Hemizygous mice are viable and fertile, exhibiting attenuated weight gain and a progressive neurological phenotype. The neurological phenotype is characterized by prominent gait abnormalities (~4 weeks), mild tremor, moderate hypoactivity, forelimb/hindlimb clasping (~24 weeks), inability to correct during negative geotaxis (~20 weeks), marked neuronal degeneration and mild gliosis of the dentate and pontine nerve nuclei, atrophy of the cerebellar Purkinje and molecular cell layers, and increased neuronal intranuclear inclusions (NIIs) that are ubiquitinated. With age, mice develop deficits in beam walking and gait walking assays, as well as neuronal loss in pontine nuclei and substantia nigra regions.

Homozygous mice (MJD84.2/84.2) exhibit increased severity and earlier onset of symptoms, as well as excessive grooming.

For these MJD84.2 mice, the donating investigator did not sequence the CAG repeat expansion over time. It is unknown if the CAG repeat number is subject to germline and somatic instability (whether it may expand or contract). When using lines with CAG repeat length, it is strongly recommended the CAG repeat number be quantified in all the experimental animals; all animals in all experimental groups should carry comparable CAG repeat sizes. CAG repeat sizing in mice should be done using high-resolution methods; as assays based on agarose gel electrophoresis may not provide sufficient resolution to accurately measure CAG repeat numbers. If labs do not have access to the appropriate equipment for determining CAG repeat length, CAG repeats can be evaluated on a fee-for-service basis by Laragen, Inc.

It is the experience of The Jackson Laboratory that the CAG repeat values of hemizygous MJD84.2 mice (a single genomic integration of two copies of the transgene) have been generally consistent over time, with the Laragen assay reporting ~65-78 CAG repeats [June 2020].

Development

Expression Data

Control Suggestions

Selected References

Genetics

Tg(ATXN3*)84.2Cce

Disease/Phenotype

Disease Terms

Research Areas By Phenotype

Mammalian Phenotype Terms by Genotype

References

Technical Support

CONTACT TECHNICAL SUPPORT

Genotyping Protocols

Standard PCR:Laragen
End Point Analysis:Tg(ATXN3*)84.2Cce-presence/absence
Genotyping resources and troubleshooting

Breeding Considerations

When maintaining a live colony, hemizygous mice may be bred to wildtype (noncarrier) mice from the colony or with C57BL/6J inbred mice (Stock No. 000664). Hemizygous mice exhibit a progressive neurological phenotype. Homozygous mice exhibit increased severity and earlier onset of neurological symptoms.

Additional Breeding and Husbandry Support

Mating System

Noncarrier x Hemizygote
Hemizygote x Noncarrier

Appearance
Black

Citation
When using the B6;CBA-Tg(ATXN3*)84.2Cce/JbezJ mouse strain in a publication, please cite the originating article(s) and include JAX stock #012705 in your Materials and Methods section.

Animal Health Reports
Facility Barrier Level Descriptions

AX10 (Standard)

Pricing & Availability

Available

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

Domestic | **International**

Pricing effective for USA, Canada and Mexico shipping destinations

LIVE MOUSE

AGE	SEX	GENOTYPE	PRICE
4 weeks	Female	Hemizygous for Tg(ATXN3*)84.2Cce	$255.00
	Male	Hemizygous for Tg(ATXN3*)84.2Cce	$255.00
4 weeks	Female	Noncarrier	$78.51
	Male	Noncarrier	$78.51
5 weeks	Female	Hemizygous for Tg(ATXN3*)84.2Cce	$255.00
	Male	Hemizygous for Tg(ATXN3*)84.2Cce	$255.00
5 weeks	Female	Noncarrier	$78.51
	Male	Noncarrier	$78.51
6 weeks	Female	Hemizygous for Tg(ATXN3*)84.2Cce	$255.00
	Male	Hemizygous for Tg(ATXN3*)84.2Cce	$255.00
6 weeks	Female	Noncarrier	$78.51
	Male	Noncarrier	$78.51
7 weeks	Female	Hemizygous for Tg(ATXN3*)84.2Cce	$255.00
	Male	Hemizygous for Tg(ATXN3*)84.2Cce	$255.00
7 weeks	Female	Noncarrier	$78.51
	Male	Noncarrier	$78.51
8 weeks	Female	Hemizygous for Tg(ATXN3*)84.2Cce	$255.00
	Male	Hemizygous for Tg(ATXN3*)84.2Cce	$255.00

	SEX		
8 weeks		Noncarrier	$78.51
	Male	Noncarrier	$78.51
9 weeks	Female	Hemizygous for Tg(ATXN3*)84.2Cce	$255.00
	Male	Hemizygous for Tg(ATXN3*)84.2Cce	$255.00
9 weeks	Female	Noncarrier	$78.51
	Male	Noncarrier	$78.51
10 weeks	Female	Hemizygous for Tg(ATXN3*)84.2Cce	$255.00
	Male	Hemizygous for Tg(ATXN3*)84.2Cce	$255.00
10 weeks	Female	Noncarrier	$78.51
	Male	Noncarrier	$78.51
11 weeks	Female	Hemizygous for Tg(ATXN3*)84.2Cce	$255.00
	Male	Hemizygous for Tg(ATXN3*)84.2Cce	$255.00
11 weeks	Female	Noncarrier	$78.51
	Male	Noncarrier	$78.51
12 weeks	Female	Hemizygous for Tg(ATXN3*)84.2Cce	$255.00
	Male	Hemizygous for Tg(ATXN3*)84.2Cce	$255.00
12 weeks	Female	Noncarrier	$78.51
	Male	Noncarrier	$78.51

BREEDER PAIR

SEX	GENOTYPE	PRICE
Female	Hemizygous for Tg(ATXN3*)84.2Cce	$333.51
Male	Noncarrier	
Female	Noncarrier	$333.51
Male	Hemizygous for Tg(ATXN3*)84.2Cce	

CRYORECOVERY - DOMESTIC PRICING

SERVICE/PRODUCT	DESCRIPTION	PRICE
Cryo Recovery	Hemizygous or non-carrier for Tg(ATXN3*)84.2Cce	$2,854.50

RELATED PRODUCTS AND SERVICES

Frozen Mouse Embryo	B6;CBA-Tg(ATXN3*)84.2Cce/IbezJ Frozen Embryo	$2595.00

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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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. We do not guarantee breeding performance and therefore suggest that investigators order more than one breeding pair to avoid delays in their research.

Terms Of Use

TERMS OF USE

General Terms and Conditions

QUESTIONS ABOUT TERMS OF USE

ADDITIONAL USE RESTRICTIONS APPLY

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LICENSING INFORMATION

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

Related Strains

All

By Allele

By Gene

By Collection