The Jackson Laboratory

# C57BL/6J-Tg(LMNB1)1Yfu/J

Stock No: **023083**

Coisogenic, Transgenic

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

PLACE ORDER

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impairment and epilepsy, followed by age-dependent motor deficits.

## Donating Investigator

Ying-Hui Fu, University of California, San Francisco

Louis Ptacek, University of California, San Francisco

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## GENETIC OVERVIEW

**Genetic Background** **Generation**

### Tg(Lmnb1)1Yfu

**Allele Type**

Transgenic (Inserted expressed sequence)

VIEW GENETICS

## RESEARCH APPLICATIONS

Neurobiology Research

VIEW ALL RESEARCH APPLICATIO

BASE PRICE

Starting at:

$2,854.50 Domestic price Cryo Recovery

VIEW PRICE LIST

## Details

### Detailed Description

Adult-onset autosomal-dominant leukodystrophy (ADLD) is a progressive and fatal neurological disorder characterized by early autonomic dysfunction, cognitive impairment, pyramidal tract and cerebellar dysfunction, and white matter loss in the central nervous system. ADLD is caused by duplication of the *LMNB1* gene, which results in increased lamin B1 transcripts and protein expression.

This transgenic model of ADLD expresses four times the normal levels of full-length mouse *Lmnb1* under the control of the endogenous promoter and regulatory sequences. Expression levels are approximately 4-fold higher compared with wildtype.

Animals are born healthy and are overtly indistinguishable from control littermates, but eventually recapitulate many of the features of ADLD. Mice exhibit cognitive impairment and epilepsy, followed by age-dependent motor deficits.

Substantial spatial memory deficits are observed at 12 months of age in a Morris water maze assay and exhibit impairments in a step-through passive avoidance task, which reflects long-term spatial associative learning and fear memory of an aversive experience. Transgenic animals exhibit progressive motor impairment on 2 different motor tasks, the accelerated rotarod (at 24 months of age) and balance beam (progressing at 12 months).

Frequent behavioral seizures, varying from generalized spasms and tremors to generalized clonic activity with atonia and tail extension are seen. Cortical EEG reveals frequent spontaneous epileptic activity, including epileptiform discharges and seizures. Transgenic mice exhibit an approximately 20-fold increase in the number of interictal spikes. Overexpression results in aberrant myelin formation and axonal degeneration (at 12 months of age), and demyelination (by 24 months).

### Development

### Expression Data

### Control Suggestions

### Selected References

## Genetics

### Tg(Lmnb1)1Yfu

## Disease/Phenotype

### Disease Terms

### Research Areas By Phenotype

### Mammalian Phenotype Terms by Genotype

### References

## Technical Support

CONTACT TECHNICAL SUPPORT

### Genotyping Protocols

Standard PCR:Tg(LMNB1)1Yfu-5'
Genotyping resources and troubleshooting

### Breeding Considerations

Hemizygotes are viable and fertile, but homozygotes are not.

Additional Breeding and Husbandry Support

### Citation

When using the C57BL/6J-Tg(LMNB1)1Yfu/J mouse strain in a publication, please cite the originating article(s) and include JAX stock #023083 in your Materials and Methods section.

### Animal Health Reports

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

CRYORECOVERY - DOMESTIC PRICING

| SERVICE/PRODUCT | DESCRIPTION | PRICE |
|---|---|---|
| Cryo Recovery | Hemizygous or Non Carrier for Tg(LMNB1)1Yfu | $2,854.50 |

RELATED PRODUCTS AND SERVICES

| | | |
|---|---|---|
| Frozen Mouse Embryo | C57BL/6J-Tg(LMNB1)1Yfu/J 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.

### THE JACKSON LABORATORY'S GENOTYPE PROMISE

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

Use of MICE by companies or for-profit entities requires a license prior to shipping.

## LICENSING INFORMATION

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

## Related Strains

All

By Allele

By Gene

By Collection