# B6.129(Cg)-*Cln3* $^{tm1.1Mem}$/J

Stock No: **017895** | Cln3deltaex7/8

Congenic, Targeted Mutation

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

PLACE ORDER



and accumulate mitochondrial ATPase subunit c, and may be useful in studies of juvenile onset neuronal ceroid lipofuscinosis (JNCL), also known as Batten or Spielmeyer-Vogt disease.

## Donating Investigator

Susan Cotman, Massachusetts General Hospital

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## GENETIC OVERVIEW

| Genetic Background | Generation |
|---|---|

### *Cln3*$^{tm1.1Mem}$

| Allele Type | Gene Symbol | Gene Name |
|---|---|---|
| Targeted (Null/Knockout) | *Cln3* | ceroid lipofuscinosis, neuronal 3, juvenile (Batten, Spielmeyer-Vogt disease) |

VIEW GENETICS

## RESEARCH APPLICATIONS

Neurobiology Research
Research Tools
Sensorineural Research

VIEW ALL RESEARCH APPLICATIO

BASE PRICE

Starting at:

$2,854.50 Domestic price Cryo Recovery

VIEW PRICE LIST

## Details

### Detailed Description

These mice carry a 1.02kb deletion mutation that recapitulates the most common mutation (>80%) found in juvenile-onset neuronal ceroid lipofuscinosis (JNCL) patients.
Mice that are homozygous for the targeted mutation on the C57BL/6 background are viable and fertile. Northern blot and RT PCR analyses of kidney, liver and brain tissue from homozygotes detect mutant mRNA. Truncated polypeptide and non-truncated alternatively spliced gene products are present. Autofluorescent lysosomal material containing immunoreactive ATPase subunit c are found in the brain and heart. Homozygous mice on the C57BL/6N background exhibit progressive retinal degeneration starting at 20 weeks of age. At 10-14 weeks of age homozygotes display slight deficits in sensory and motor tasks. Increased rectal body temperature and minimum oxygen consumption are observed in homozygotes 12-13 weeks of age. The heart weight of 20 week old homozygotes is slightly increased, although cardiac function is normal in young adults. At 15-16 weeks of age homozygotes on the C57BL/6N background exhibit exhibit elevated serum ferritin levels, mean red blood cell corpuscular volume, and reticulocyte numbers. Homozygotes have vacuolated peripheral blood lymphocytes (observed as early as the neonatal period), and male homozygotes exhibit vacuolation in epididymal clear cells.

### Development

### Control Suggestions

### Selected References

## Genetics

### *Cln3*$^{tm1.1Mem}$

## Disease/Phenotype

Disease Terms

Research Areas By Phenotype

Mammalian Phenotype Terms by Genotype

References

## Technical Support

CONTACT TECHNICAL SUPPORT

### Genotyping Protocols
Standard PCR:Cln3alternate2
Genotyping resources and troubleshooting

### Breeding Considerations

When maintaining a live colony, these mice can be bred as homozygotes.

Additional Breeding and Husbandry Support

### Citation
When using the Cln3deltaex7/8 mouse strain in a publication, please cite the originating article(s) and include JAX stock #017895 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 | Heterozygous or wildtype for Cln3<tm1.1Mem> | $2,854.50 |

| RELATED PRODUCTS AND SERVICES | | |
|---|---|---|
| Frozen Mouse Embryo | B6.129(Cg)-Cln3<tm1.1Mem>/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

TOMORROW'S CURES



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