The Jackson Laboratory

B6.Cg-*Usp14*$^{ax\text{-}J}$/J

Stock No: **000518** | ataxia Jackson

Congenic

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

PLACE ORDER

Email Download PDF Help

READ MORE +

GENETIC OVERVIEW

Genetic Background	Generation

Usp14$^{ax\text{-}J}$

Allele Type	Gene Symbol	Gene Name
Spontaneous (Hypomorph)	*Usp14*	ubiquitin specific peptidase 14

VIEW GENETICS

RESEARCH APPLICATIONS

Apoptosis Research
Developmental Biology Research
Neurobiology Research
Cell Biology Research

VIEW ALL RESEARCH APPLICATIO

BASE PRICE

Starting at:

$2,854.50 Domestic price Cryo Recovery

VIEW PRICE LIST

Details

Detailed Description

The first outward sign of homozygosity for the recessive mutation *Usp14*$^{ax\text{-}J}$ is an unsteady gait, particularly in the hind quarters, that can be detected by approximately 2 weeks of age. These pups often show difficulty in righting themselves when turned on their backs. They have a rapid tremor that is most apparent when they are active. Extensor paralysis progresses and wasting of the hind quarters ensues until the hind quarters are not functional and the mouse can not sit upright or move except through the use of onlyits front limbs. Although death is premature, viability in the first two to three weeks is not diminished. At 18 days of age these mutants weigh an average of 15% less than wildtype controls and by 45 days this difference is increased such that they weigh 50-60% less than wildtype controls. Homozygotes do not breed. The lumbar vertebrae are slightly shorter than normal with shorter spinous processes and more elongate foramen. These homozygotes have defects in synaptic transmission in the central and peripheral nervous systems. End plate potentials are altered at neuromuscular junctions. D'Amato et al. published an extensive histological assessment of the *Usp14*$^{ax\text{-}J}$/*Usp14*$^{ax\text{-}J}$ brain in which they reported that the corpus collosum, cingulum, and hippocampal commissureare underdeveloped as are particular nuclei and long tracts of the brain stem. The pons is shorter longitudinally and deficiencies are also found in the trapezoid body, as well as spinal nerves and spinal cord white matter. The medial lemniscus is smaller than normal only from the ventral beginning in the medulla to an area rostral to the substantia nigra and cerebral peduncles (for more detail see D'Amato et al., 1965.). A.M. Burt described abnormalities in the differentiation of the dendritic trees of CA1 pyramidal cells of the hippocampus and granule cells of the dentate gyrus. This is consistent with altered developmental apoptosis. Ohgoh et al. subsequently found significant apoptosis in the granule cell layer of the cerebellum but not the dentate gyrus or olfactory bulb. Ohgoh et al. also found no TUNEL staining of Purkinje cells, but the early report by D'Amato et al. identified disordered development and degenerative deformities of Purkinje cells. (Lyon, 1955; D'Amato and Hicks, 1965; Burt, 1980; Ohgoh et al., 2000; Wilson et al., 2002.)

Development

Control Suggestions

Genetics

Usp14$^{ax\text{-}J}$

Disease/Phenotype

Disease Terms

Research Areas By Phenotype

Mammalian Phenotype Terms by Genotype

References

Technical Support

CONTACT TECHNICAL SUPPORT

Genotyping Protocols

Standard PCR:Usp14
Genotyping resources and troubleshooting

Citation

When using the ataxia Jackson mouse strain in a publication, please cite the originating article(s) and include JAX stock #000518 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 for Usp14<ax-J>	$2,854.50

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

LICENSING INFORMATION

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

Related Strains

All

By Allele

By Gene

By Collection

▲