The Jackson Laboratory

129S(Cg)-*Tgfbr2* $^{tm1.1Hcd}$ /J

Stock No: **024634**

Congenic, Targeted Mutation

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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Harry Dietz, Johns Hopkins Medical Institute

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GENETIC OVERVIEW

Genetic Background	Generation

Tgfbr2$^{tm1.1Hcd}$

Allele Type	Gene Symbol	Gene Name
Targeted	*Tgfbr2*	transforming growth factor, beta receptor II

VIEW GENETICS

RESEARCH APPLICATIONS

Internal/Organ Research
Cardiovascular Research
Developmental Biology Research

VIEW ALL RESEARCH APPLICATIO

BASE PRICE

Starting at:

$2,854.50 Domestic price Cryo Recovery

VIEW PRICE LIST

Details

Detailed Description

Loeys-Dietz Syndrome (LDS) is a connective tissue disorder that is characterized by a high risk for aneurysm and dissection throughout the arterial tree and phenotypically resembles Marfan Syndrome. LDS is caused by heterozygous missense mutations in either TGF receptor gene (*TGFBR1* or *TGFBR2*).

This targeted mutant strain carries an G357W missense mutation associated with LDS in the mouse *Tgfbr2* (transforming growth factor, beta receptor II) gene. Mutant and wildtype alleles are expressed approximately equally in the aortic walls of heterozygotes. Heterozygous mice develop progressive aortic root aneurysm, as well as elongation and tortuosity of the aortic arch and coronary arteries. A subtle but significant deviation from the wildtype condition is detectable at 4 weeks of age, and becomes highly reproducible and dramatic at 24 weeks. At 24 weeks of age, elastic fiber fragmentation is detectable in the aortic roots of these knockin mice. The aortic wall also shows progressive thickening with excessive collagen deposition. Approximately 10-15% of heterozygotes may die by 180 days of age due to aortic dissection. Hemothorax or hemopericardium is observable in approximately 60% of deaths. These mice also develop skeletal manifestations associated with LDS, such as kyphosis, overgrowth of the ribs and craniosynostosis.

Development

Control Suggestions

Selected References

Genetics

Tgfbr2$^{tm1.1Hcd}$

Disease/Phenotype

Disease Terms

Research Areas By Phenotype

Mammalian Phenotype Terms by Genotype

References

Technical Support

CONTACT TECHNICAL SUPPORT

Genotyping Protocols
Standard PCR:Tgfbr2
Standard PCR:Generic Neo
Probe:Generic Neo
Genotyping resources and troubleshooting

Breeding Considerations

Heterozygotes are fertile, but show reduced viability. Approximately 10-15% of heterozygotes may die by 180 days of age due to aortic dissection. Homozygotes are not viable.

Additional Breeding and Husbandry Support

Citation
When using the 129S(Cg)-*Tgfbr2*$^{tm1.1Hcd}$/J mouse strain in a publication, please cite the originating article(s) and include JAX stock #024634 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 **Internatic**

Pricing effective for USA, Canada and Mexico shipping destinations

CRYORECOVERY - DOMESTIC PRICING

SERVICE/PRODUCT	DESCRIPTION	PRICE
Cryo Recovery	Heterozygous or Wilt type for Tgfbr2<tm1.1Hcd>	$2,854.50

RELATED PRODUCTS AND SERVICES

Frozen Mouse Embryo	129S(Cg)-Tgfbr2<tm1.1Hcd>/J Frozen Embryo	$2595.00

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Terms Of Use

TERMS OF USE

General Terms and Conditions

QUESTIONS ABOUT TERMS OF USE

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LICENSING INFORMATION

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

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All

By Allele

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