

STOCK *Fbn1*^{tm3.1Hcd}/J

Stock No: 025474

 Targeted Mutation

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

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topoisomerase I antibodies.

Donating Investigator

Harry Dietz, Johns Hopkins Medical Institute

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GENETIC OVERVIEW

Genetic Background

Generation

Fbn1^{tm3.1Hcd}

Alele Type

Targeted (Humanized sequence)

Gene Symbol

Fbn1

Gene Name

fibrillin 1

VIEW GENETICS

RESEARCH APPLICATIONS

Immunology, Inflammation and Autoimmunity Research

Dermatology Research

Developmental Biology Research

VIEW ALL RESEARCH APPLICATIONS

BASE PRICE

Starting at:

\$2,854.50 Domestic price Cryo Recovery

V I E W P R I C E L I S T

Details

Detailed Description

Stiff skin syndrome (SSS), a form of scleroderma defined by a pathological fibrosis of the skin, is caused by heterozygous missense mutations in the Arg-Gly-Asp (RGD) integrin-binding domain of the *FBN1* (fibrillin 1) gene.

These mice carry a W1572C mutation (equivalent of the human W1570C mutation) in the mouse *Fbn1* gene. The mutation is widely expressed in tissues including the skin, aorta, lungs and liver. Heterozygotes serve as a model of human stiff skin syndrome with increased collagen deposition in the dermis by 1 month of age, decreased subcutaneous fat by 3 months of age, disorganized and excessive microfibrillar aggregates in the dermis, and circulating anti-nuclear and anti-topoisomerase I antibodies by three months of age. Mice show skin infiltration of pro-inflammatory immune cells including plasmacytoid dendritic cells, T helper cells, and plasma cells. Mice homozygous for the W1572C mutation are viable and show accelerated skin fibrosis when compared with heterozygous littermates.

Development

Control Suggestions

Selected References

Genetics

Fbn1^{tm3.1Hcd}

Disease/Phenotype

Disease Terms

[+ Research Areas By Phenotype](#)

[+ Mammalian Phenotype Terms by Genotype](#)

[+ References](#)

[- Technical Support](#)

C O N T A C T T E C H N I C A L S U P P O R T

Genotyping Protocols

Sanger sequencing:[Fbn1alternate1](#)

End Point Analysis:[Fbn1 EP](#)

[Genotyping resources and troubleshooting](#)

Breeding Considerations

Heterozygotes are viable and fertile. Homozygous fertility is reduced slightly in males, more so in females.

[Additional Breeding and Husbandry Support](#)

Citation

When using the STOCK *Fbn1*^{tm3.1Hcd}/J mouse strain in a publication, please [cite the originating article\(s\)](#) and include JAX stock #025474 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 Fbn1<tm3.1Hcd>	\$2,854.50

RELATED PRODUCTS AND SERVICES		
Frozen Mouse Embryo	STOCK Fbn1<tm3.1Hcd>/J	\$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

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[General Terms and Conditions](#)

Q U E S T I O N S A B O U T T E R M S O F U S E

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



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