B6.Cg-Fbn1<sup>Tsk</sup>/J

Stock No: 014632 | B6-tight skin

- Congenic Spontaneous Mutation

**REPOSITORY LIVE**

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**PLACE ORDER**

3-6 week lead time for most orders depending on quantity and age range requested

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**Overview**

**Also Known As: B6-tight skin**

Fbn1<sup>Tsk</sup> homozygotes are embryonic lethal; heterozygotes exhibit excessive growth and hyperplasia of connective tissue, cartilage, tendon sheaths and bone. This strain is a model for Marfan Syndrome, Hereditary Pulmonary Emphysema, Stiff Skin Syndrome and Familial Progressive Scleroderma.

**Donating Investigator**

IMR Colony, The Jackson Laboratory

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**GENETIC OVERVIEW**

<table>
<thead>
<tr>
<th>Genetic Background</th>
<th>Generation</th>
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RESEARCH APPLICATIONS
Internal/Organ Research
Mouse/Human Gene Homologs
Dermatology Research
Developmental Biology Research
Immunology, Inflammation and Autoimmunity Research
Research Tools
Cell Biology Research
Cardiovascular Research

BASE PRICE
Starting at:
$76.22 Domestic price for female
257.05 Domestic price for breeder pair

Details

Detailed Description
The Fbn1<sup>Tsk</sup> allele contains a 30 to 40 kbp genomic tandem duplication resulting in a larger than normal in-frame transcript. Heterozygotes are embryonic lethal, failing to survive past somite formation (7-8 days of gestation). Heterozygotes are viable and fertile, exhibiting excessive growth and hyperplasia of connective tissue, cartilage, tendon sheaths and bone. Skin tightness, due to hyperplastic thickening of subcutaneous loose connective tissue and abnormal organization and distribution of skin microfibrillar arrays, develops by the first week after birth. Although the size of the skeleton is increased, body weight remains normal. Mutant mice exhibit polyuria during the light cycle. Collagens and glycosaminoglycans accumulate in the skin, heart, lungs and bladder. Hypertrophy is also observed in the enlarged heart (aortic adventitia). Mutant mice have enlarged thoracic size and lungs with abnormal alveolar walls, irregular shaped alveoli, and increased lung capacity. By 1 month of age, heterozygotes develop emphysema with progressive destruction of alveolar walls and loss of elasticity. Inflammatory macrophages and neutrophils infiltrate the lower respiratory tract. The abnormal immune system characteristics exhibited by this strain also include: alveolitis, increased number of mast cells in the skin, presence of autoantibodies specific for scleroderma target antigens and anti-nuclear antibodies.

In an attempt to offer alleles on well-characterized or multiple genetic backgrounds, alleles are frequently moved to a genetic background different from that on which an allele was first characterized. This is the case for the strain above. It should be noted that
the phenotype could vary from that originally described. We will modify the strain description if necessary as published results become available.

Genotyping Protocols
Standard PCR: Fbn1^Ts

Genotyping resources and troubleshooting

Dietary Information
LabDiet® 5K52 formulation (6% fat)

Breeding Considerations
Homozygotes are not viable. When maintaining a live colony, C57BL/6J female mice may be bred with heterozygous male mice. Additional Breeding and Husbandry Support

Mating System
C57BL/6J (000664) x Heterozygote

Appearance
Hair loss has been observed in this colony.

Citation
When using the Fbn1^Ts mouse strain in a publication, please cite the originating article(s) and include JAX stock #014632 in your Materials and Methods section.

Facility Barrier Level Descriptions
**Pricing & Availability**

3–6 week lead time for most orders depending on quantity and age range requested

### Domestic | International

**Pricing effective for USA, Canada and Mexico shipping destinations**

#### Live Mouse

<table>
<thead>
<tr>
<th>AGE</th>
<th>SEX</th>
<th>GENOTYPE</th>
<th>PRICE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Approx 4-8 weeks</td>
<td>Female</td>
<td>Heterozygous for Fbn1&lt;sup&gt;Tsk&lt;/sup&gt;</td>
<td>$231.00</td>
</tr>
<tr>
<td></td>
<td>Male</td>
<td>Heterozygous for Fbn1&lt;sup&gt;Tsk&lt;/sup&gt;</td>
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<td>Wild-type for Fbn1&lt;sup&gt;Tsk&lt;/sup&gt;</td>
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<td>$76.22</td>
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</tbody>
</table>

#### Breeder Pair

<table>
<thead>
<tr>
<th>SEX</th>
<th>GENOTYPE</th>
<th>PRICE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Female</td>
<td>C57BL/6J (000664)</td>
<td>$257.05</td>
</tr>
<tr>
<td>Male</td>
<td>Heterozygous for Fbn1&lt;sup&gt;Tsk&lt;/sup&gt;</td>
<td></td>
</tr>
</tbody>
</table>

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Terms of Use

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Phone: 207-288-6470

Email: TechTran@jax.org

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