RB156Bnr/Ei-Dsp<sup>nu1</sup>/GrsrJ

Stock No: 005362 | ruffled

Coisogenic, Spontaneous Mutation

CRYO RECOVERY

PLACE ORDER

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

Also Known As: ruffled

This spontaneous C-terminal truncation provides a uniquely viable mouse model for Carvajal-Huerta syndrome, with defective desmosomes resulting in abnormal fur, epidermal blistering, follicular dystrophy of the vibrissa, and ventricular fibrosis with electrocardiographic abnormalities.

READ MORE »

GENETIC OVERVIEW

<table>
<thead>
<tr>
<th>Genetic Background</th>
<th>Generation</th>
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<tbody>
<tr>
<td>Dsp&lt;sup&gt;nu1&lt;/sup&gt;</td>
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<table>
<thead>
<tr>
<th>Allele Type</th>
<th>Gene Symbol</th>
<th>Gene Name</th>
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<tbody>
<tr>
<td>Spontaneous (Hypomorph)</td>
<td>Dsp</td>
<td>desmoplakin</td>
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Marker(s)

<table>
<thead>
<tr>
<th>Marker Symbol</th>
<th>Marker Name</th>
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<tbody>
<tr>
<td>Rb(9.14)6Bnr</td>
<td>Robertsonian translocation, Chr 9 and 14, Universitat Bonn/Rhein 6</td>
</tr>
<tr>
<td>Rb(8.12)5Bnr</td>
<td>Robertsonian translocation, Chr 8 and 12, Universitat Bonn/Rhein 5</td>
</tr>
<tr>
<td>Rb(1.3)1Bnr</td>
<td>Robertsonian translocation, Chr 1 and 3, Universitat Bonn/Rhein 1</td>
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</tbody>
</table>

VIEW GENETICS

RESEARCH APPLICATIONS
Dermatology Research
Developmental Biology Research
Internal/Organel Research
Cardiovascular Research
## BASE PRICE
Starting at:

$2,595.00 Domestic price Cryo Recovery

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## Important Note
This genetic background is homozygous for Robertsonian translocations Rb(1.3)18nr, Rb(8.12)58nr, and Rb(9.14)68nr.

## Detailed Description
The phenotype of this mutation can be identified at about 10 days of age by a ruffled looking coat, which persists throughout the animal's lifespan, unlike the caracul and mutants, which lose some of their curly coat with age. The hair of ruffled homozygotes appears disheveled, slightly sparse, and has some curling. Horizontal sections show hair shafts are larger in diameter than normal, with loss of concentric layering, and vertical sections show loss of the septation pattern of the medulla and a pointed tip. Although the pelage hair follicles appear normal there is widespread follicular dystrophy of the vibrissae. Dorsal skin sections show acanthosis and the mouth and tongue have suprabasilar blistering and pustule formation. Fibrosis develops with age in the left ventricular free wall and apex of the heart and electrocardiograms at 4 weeks of age show longer RR and GGR intervals, smaller R wave and T wave amplitudes and slower heart rate, suggestive of a form of atrial tachycardia. Heterozygotes also show some electrocardiographic defects with age. Both male and female homozygotes breed normally. Unlike null alleles of desmoplakin, which are embryonic lethal, this spontaneous C-terminal truncation provides a novel viable model for Carvajal-Huerta syndrome.

## Development

## Control Suggestions

## Selected References

## Genetics

### Dsp<sup>nr</sup>

### Rb(9.14)68nr

### Rb(8.12)58nr

### Rb(1.3)18nr

## Disease/Phenotype
Genotyping Protocols
Genotyping resources and troubleshooting

Citation
When using this NIH-funded mouse strain in a publication, please cite the originating article(s) and include JAX stock #005362 in your Materials and Methods section.
Production of mice from cryopreserved embryos or sperm occurs in a maximum barrier room, G200

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

<table>
<thead>
<tr>
<th>Domestic</th>
<th>International</th>
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<tr>
<td>Pricing effective for USA, Canada and Mexico shipping destinations</td>
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Cryorecovery - Domestic Pricing

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<tr>
<th>SERVICE</th>
<th>GENOTYPE</th>
<th>PRICE</th>
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<tbody>
<tr>
<td>Cryo Recovery</td>
<td>Heterozygous for rul</td>
<td>$2,595.00</td>
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We will fulfill your order by providing at least two carriers for each strain ordered. The total number, sex, and genotypes provided will vary, although typically 8 or more animals are provided. Please check genotypes which will be recovered. While the genotypes of all animals produced will be communicated to you prior to scheduling shipment, the genotypes of animals provided may not reflect the mating scheme and genotypes described in the strain description. Animals are typically ready to ship in 11-14 weeks. If a second recovery is required to produce the minimum number of animals, then delivery time would increase to approximately 25 weeks. If we fail to produce animals of the correct genotype, you will not be charged. We cannot guarantee the reproductive success of mice.
shipped to your facility. If the mice are lost after the first three days (post-arrival) or do not produce progeny at your facility, a new order and fee will be necessary.

Cryorecovery to establish a Dedicated Supply for greater quantities of mice. Mice recovered can be used to establish a dedicated colony to contractually supply you mice according to your requirements. Price by quotation.

### Related Products and Services

| Frozen Mouse Embryo | $2,595.00 per straw or vial |

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<td>By Collection</td>
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All Related Strains