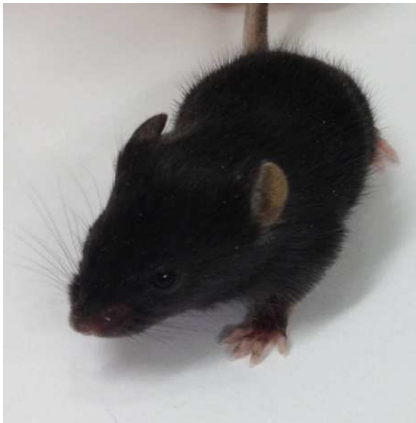


## SCID/BLAJ MOUSE

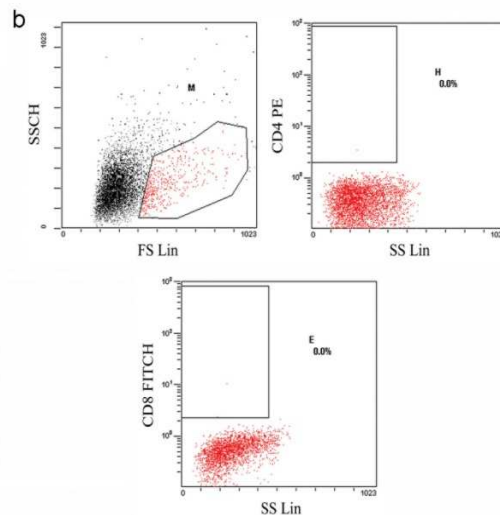
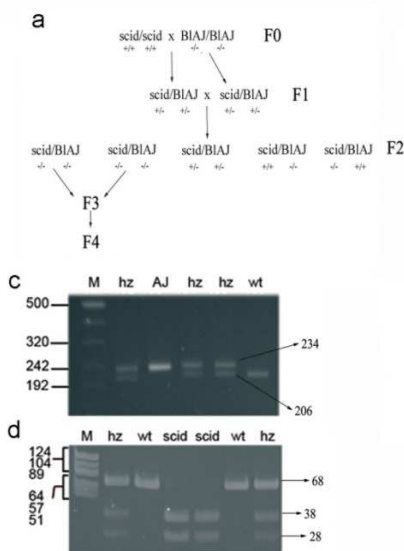


| SCID/BLAJ MOUSE |   |
|-----------------|---|
| CODE            | ramY_BIAJ   |
| BREEDING        | A cross between CB17/lcr-Prkdc <sup>scid</sup> /lcrCrl and BIAJ |
| COAT COLOR      | Brown   |

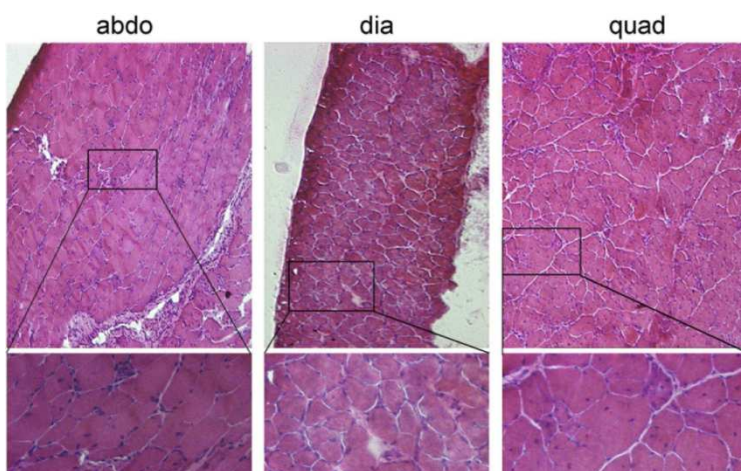
### RESEARCH APPLICATION

Scid/BLAJ represents a suitable model for preclinical studies concerning stem cell transplantation.

### DESCRIPTION



BLAJ mice were bred to the *C.B-17/lcr scid/scid* mice. The resulting F1 (*Scid/blaj +/-*) mice have been crossed to obtain F2 homozygotes for both loci: *dysf* and *Scid*. Two additional generations of *Scid/blaj -/-* have been performed, until F4. CD8<sup>+</sup> or CD4<sup>+</sup> cells were not found in the blood of *Scid/blaj* mice. BIAJ mice have been characterized for the presence of specific ETn retrotransposon. In wild type mice, the length of DNA fragment for dysferlin is 206-bp. In heterozygous mice, two bands of 242-bp and 206-bp have been obtained, by using the primers for dysferlin with one specific for the ETn retrotransposon. Homozygous mice have been characterized by only one band of 242-bp. Normal mice are characterized by only one 68-bp DNA fragment. In homozygous mice, 2 fragments of 28-bp and 38 bp have been obtained while in heterozygous ones 3 fragments of 68-bp, 38-bp and 28-bp have been reported.



Scid/BLAJ muscles show the presence of degenerating and small centrally-nucleated regenerating fibers. These dystrophic changes are primarily found in the quadriceps (QA), abdominal (ABDO) and diaphragm (DIA) muscles whereas gastrocnemius, soleus and tibialis anterior are unaffected. By 9 months of age, there are active myopathy of different severity in all the skeletal muscles examined and hypertrophic fibers, fiber splitting and fat replacement are also evident. In *scid/blaj* mice, ABDO and DIA muscles are characterized by increased numbers of necrotic and regenerating fibers, infiltration of mononuclear cells into intact fibers, phagocytosis and marked variation of fiber size.

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