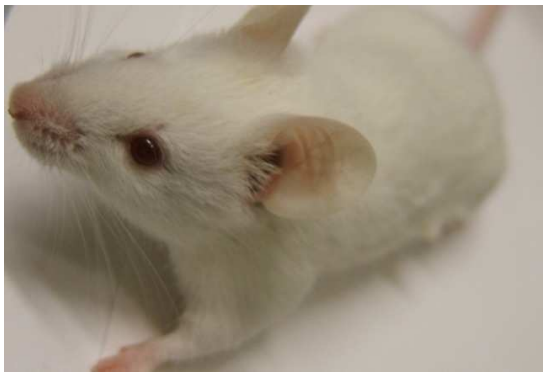
MDX/SCID MOUSE



MDX/SCID MOUSE	
CODE	ramY_mdx
BREEDING	A cross between CB17/Icr-*Prkdc*^{scid}/IcrCrl and C57BL/10ScSn-*Dmd*^{mdx}/J
COAT COLOR	White

RESEARCH APPLICATION

Mdx/scid mouse represents a suitable model for preclinical studies concerning stem cell transplantation.

DESCRIPTION

Scid/Mdx mouse is an immunodepressed dystrophic animal model. Peripheral blood of littermates or age-matched double homozygous scid/mdx mice was analyzed for the presence of B and T lymphocytes expressing CD4, CD8, CD19 and B220. In the double homozygous scid/mdx mice, a mean percentage of 9.3% of total blood-derived cells expressing B220 antigen (n = 100), while no CD4+, CD8+ or CD19+ cells has been found. To genotype the dystrophin gene, PCR analysis of DNA from the scid/mdx mice has been performed after digestion with Mae III restriction enzyme (endonuclease). Using this method, we have distinguished mice heterozygous and homozygous at the dystrophin locus throughout the F1–F12 progeny mice on the basis of the detection of four (153 bp, 48 bp, 26 bp and 22 bp) or only two (153 bp and 48 bp) products, respectively.

In the *scid*/*mdx* mice, centrally nucleated fibres with different diameters (H&E) and fibrosis among the muscle fibres (blue in Azan Mallory) are present, which suggests that the *scid/mdx* muscles have a dystrophic phenotype. The proportion of *scid*/*mdx* muscle fibres with centrally located nuclei was 46–52%. In *scid*/*mdx* mice, the area of the muscle fibres is 1500–1800 μm^2. In *scid*/*mdx* mice, the coefficient of variance of the muscle fibre area is 55–65.

REFERENCES

- Novel insight into stem cell trafficking in dystrophic muscles. *A. Farini, C. Villa, A. Manescu, F. Fiori, A. Giuliani, P. Razini, C. Sitzia, G. Del Fraro, M. Belicchi, M. Meregalli, F. Rustichelli, Y. Torrente.* Int J Nanomedicine. 2012;7:3059-67. Epub 2012 Jun 20.

- T and B lymphocyte depletion has a marked effect on the fibrosis of dystrophic skeletal muscles in the Scid/Mdx mouse. *A. Farini, M. Meregalli, M. Belicchi, M. Battistelli, D. Parolini, G. D'Antona, M. Gavina, L. Ottoboni,G. Constantin, R. Bottinelli and Y. Torrente.* Journal Of Pathology. 2007; 213(2):229-38

- Restoration of human dystrophin following transplantation of exon-skipping engineered DMD patient stem cells into dystrophic mice. *R. Benchaouir, M. Meregalli, A. Farini, G. D'Antona, M. Belicchi, A. Goyenvalle, M. Battistelli, N. Bresolin, R. Bottinelli, L. Garcia, and Y. Torrente.* Cell Stem Cell. 2007;1, 1-13

- High-resolution X-ray microtomography for three-dimensional visualization of human stem cell muscle homing. *Y. Torrente, M. Gavina, M. Belicchi, F. Fiori, V. Komlev, N. Bresolin, F. Rustichelli*. FEBS Lett. 2006 Oct 16;580(24):5759-64

- VCAM-1 expression on dystrophic muscle vessels has a critical role in the recruitment of human blood-derived CD133+ stem cells after intra-arterial transplantation. *M. Gavina, M. Belicchi, B. Rossi, L. Ottoboni, F. Colombo, M.Meregalli, M. Battistelli, L. Forsenigo, P. Biondetti, F. Pisati, D. Parolini, A. Farini, A.C. Issekutz, N. Bresolin, F. Rustichelli, G. Constantin and Y. Torrente.* Blood 2006 Oct 15; 108(8):2857-66

- Human Circulating AC133+ Stem Cells Replenish the Satellite Cell Pool, Restore Dystrophin Expression and Ameliorate Function Upon Transplantation in Murine Dystrophic Skeletal Muscle.*Y. Torrente, M. Belicchi, M. Sampaolesi, F. Pisati, M. Meregalli,G. D'Antona,R. Tonlorenzi,L. Porretti, M. Gavina, K. Mamchaoui, M. A. Pellegrino, D. Furling, V. Mouly, G.S. Butler-Browne, R. Bottinelli,G. Cossu, N. Bresolin.* J ClinInvest. 2004 Jul;114(2):182-95

CONTACTS

For further information, please contact Technical Services at mirella.meregalli@ystem-life.com

Ystem