Overview

Also Known As: mdx  R4-23/  C

Mice homozygous/hemizygous for the spontaneous Dmd^{mdx} allele and homozygous for the human micro-dystrophin ( R4-23/  C) transgene exhibit improved skeletal muscle function and muscle histopathology as compared to mdx mice and may be useful for studying Duchenne muscular dystrophy.
Duchenne muscular dystrophy (DMD) is a progressive muscular disorder caused by an imbalance between muscle degeneration and regeneration resulting in muscle degeneration, necrosis, accumulation of fat and fibrosis, and insufficient regeneration/loss of myofibers. The genetic cause of DMD are mutations of the dystrophin muscular dystrophy gene (DMD) on the X chromosome. The mdx mutation in mice results in a truncated DMD protein. Homozygous females and hemizygous males develop the myopathic features of DMD; although the myopathy is less severe than the human disease course. The Tg(ACTA1-DMD*)326Dua transgene carries a deletion in the human dystrophin gene of repeat 4 through repeat 23 and the C-terminus (R4-23/ C) under the control of the human skeletal alpha 1 actin (ACTA1) promoter. When the transgene is combined with the mdx allele, the donating investigator reports that homozygous mice exhibit restored skeletal muscle function and improved muscle histopathology as compared to mdx mice.
Genotyping Protocols
MELT: Tg(ACTA1-DMD*)326Dua
End Point Analysis: Dmd<sup>mdx</sup> End Point
MELT: Tg(DMD*)#Dua
Genotyping resources and troubleshooting

Breeding Considerations
While maintaining a live colony, mice carrying the X-linked mdx allele are bred as homozygote female x hemizygote male. Mice homozygous for the transgene are viable and fertile.
Additional Breeding and Husbandry Support

Citation
When using the mdx<sup>R4-23/ C</sup> mouse strain in a publication, please cite the originating article(s) and include JAX stock #027818 in your Materials and Methods section.