del52 hDMD/mdx mouse

Catalogue number: 154189
Sub-type: Mouse
Images:

Contributor

Inventor: Annemieke Aartsma-Rus
Institute: Leiden University and Leiden University Medical Center
Images:

Tool details

***FOR RESEARCH USE ONLY**

Name: del52 hDMD/mdx mouse

Alternate name: Dystrophin, Muscular Dystrophy, Duchenne And Becker Types, DXS164, DXS26, DXS23, DXS239, DXS268, DXS269, DXS27, DXS272

Class:
Conjugate:
Description: Duchenne muscular dystrophy (DMD) is a muscle-wasting disease in which muscle is continuously damaged, resulting in loss of muscle tissue and function. Antisense-mediated exon skipping is a promising therapeutic approach for DMD which uses sequence specific antisense oligonucleotides (AONs) to reframe disrupted dystrophin transcripts. As AONs function in a sequence specific manner, human specific AONs cannot be tested in the current mdx mouse model for DMD as it carries a mutation in the murine Dmd gene. In order to model the human disease more accurately we generated a mutant mouse model carrying the human DMD gene integrated in the mouse genome on an mdx background. The human DMD gene carries a deletion of exon 52 the mutation hotspot found in human disease. We also generated the control mouse for this model which carries full length human DMD (Cat No:154187) The line has NO dystrophin expression.
Purpose:
Parental cell:
Organism:
Tissue:
Model: Transgenic
Gender:
Isotype:
Reactivity:
Selectivity:
Host:
Immunogen:

Immunogen UNIPROT ID:
Sequence:
Growth properties:
Production details: The targeting vector (pGem-del52) was built on the backbone of the pGEM-T-easy vector (Promega). The targeting arms were generated by PCR using genomic DNA of the hDMD/mdx mouse ES cell line containing the complete hDMD gene as template. Intron 51 and 52 targeting arms were 3040 and 3134 bp long respectively. The negative selection marker herpes simplex virus thymidine kinase (HSV TK) was isolated from the pKO-SelectTK plasmid whereas the positive selection marker blasticidin, flanked by LoxP sites, was isolated from the pSVBsdX1 plasmid. The targeting arms and selection markers were PCR generated using the Expand Long Template PCR system of Roche. To facilitate cloning of the PCR isolated units, restriction enzyme digestion sites were added to the primers at the 5â??end. The individual units were first cloned in the pCRII-blunt TOPO vector using the Zero BluntÂŽ TOPOÂŽ PCR Cloning Kit and Sanger sequence verified prior to cloning in the destination vector. The linearized targeting construct (25 Î?g) was mixed with 50 Î?g TALEN DNA and in a 1:2 ratio mixed with LipofectamineÂŽ2000 according to the supplierâ??s manual in KO-DMEM medium without supplements. Recombinant hDMD/mdx ES cells were injected in C57BL/6J blastocysts, which were subsequently transplanted in pseudo-pregnant foster mice
Formulation:
Recommended controls:
Bacterial resistance:
Selectable markers:
Additional notes: This strain has 2 copies of the gene inserted in a tail to tail orientation for each allele. So the homozygous strain has 4 copies â?? each with an exon 52 deletion.

Target details

Target: DMD

Target alternate names:

Target background:

Molecular weight:

Ic50:

Applications

Application:
Application notes:

Handling

Format:
Concentration:

Passage number:
Growth medium:
Temperature:
Atmosphere:
Volume:
Storage medium:
Storage buffer:
Storage conditions:
Shipping conditions:

Related tools

Related tools:

References

References: Veltrop et al. 2013. PLoS Curr. 5:. PMID: 24057032