

# hDMD/mdx ES cell line

**Catalogue number:** 154188

**Sub-type:**

**Images:**

## Contributor

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**Images:**

## Tool details

**\*FOR RESEARCH USE ONLY**

**Name:** hDMD/mdx ES cell line

**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 an ES mouse cell line carrying the complete human DMD gene integrated in the mouse genome on an mdx background. This cell line was used to generate the hDMD/mdx mouse (Cat No:154187)

**Purpose:**

**Parental cell:** Blastocysts were cultured to generate ES cell lines

**Organism:** Human

**Tissue:** Embryo

**Model:** Stem Cells

**Gender:**

**Isotype:**

**Reactivity:**

**Selectivity:**

**Host:**

**Immunogen:**

**Immunogen UNIPROT ID:**

**Sequence:**

**Growth properties:**

**Production details:** Blastocysts were isolated from time mated hDMD male mice and super ovulated mdx female mice. These blastocysts were layered on murine embryonic fibroblast (MEF) feeder cells in a well of a 24-well plate, in knockout DMEM supplemented with 2 mM L-glutamine, 1 mM sodium pyruvate, non-essential amino acids, 50 units/ml of penicillin as well as streptomycin, 1000 units/ml of LIF) and 15% knockout serum replacement. Usually after 6 days blastocysts had hatched and a small colony of cells had forme...

**Formulation:**

**Recommended controls:**

**Bacterial resistance:**

**Selectable markers:**

**Additional notes:**

## Target details

**Target:** DMD

**Target alternate names:**

**Target background:**

**Molecular weight:**

**Ic50:**

## Applications

**Application:**

**Application notes:**

## Handling

**Format:** Frozen

**Concentration:**

**Passage number:**

**Growth medium:** ES medium with 15% knock serum replacement, on MEF coated plates

**Temperature:**

**Atmosphere:**

**Volume:**

**Storage medium:**

**Storage buffer:**

**Storage conditions:**

**Shipping conditions:** Dry ice

## Related tools

**Related tools:**

## References

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