



Mouse-derived FSHD muscle cell lines

Primary skeletal muscle cell lines isolated from DUX4 expressing FSHD mice.

IP Status: Research Tool

Applications

- In vitro cell culture
- Drug development

Key Benefits & Differentiators

- Derived from mouse models with DUX4 gene conformation appropriate for drug testing

Technology Overview

Facioscapulohumeral muscular dystrophy (FSHD) is one of the most frequent hereditary muscle disorders. The DUX4 gene encodes a protein that is expressed in FSHD but not in healthy muscle cells. This inappropriate expression initiates a gene deregulation cascade causing differentiation defects, muscle atrophy, and oxidative stress. To test potential drug candidates, appropriate research tools (e.g. cell lines) are needed.

Researchers at the University of Minnesota have developed primary skeletal muscle cell lines isolated from DUX4 expressing FSHD mice. These myoblast and fibroblast cell lines are appropriate for testing potential drug candidates against DUX4 protein.

Technical Information

Primary mouse myoblasts from iDUX4pA mouse model

Organism: *Mus musculus*, mouse

Tissue: Skeletal Muscle Cell Type: Myoblast

Disease: Facioscapulohumeral muscular dystrophy (FSHD)

Applications: These primary adherent cells are grown in culture to model DUX4 myopathy. Suitable for in vitro testing of drug candidates and mechanistic studies.

Product format: Frozen

Description: Primary myoblasts isolated from the skeletal muscle of FSHD mouse model described in Dandapat, A. et al. (2014). *Cell Rep.* 8(5):1484-96.

Primary mouse fibroblasts from iDUX4pA mouse model

Organism: *Mus musculus*, mouse

Tissue: Skeletal Muscle

Cell Type: Fibroblast

Technology ID

2019-199

Category

Life Sciences/Human Health
Life Sciences/Pharmaceuticals
Life Sciences/Research Tools
Life Sciences/Therapeutics

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Disease: Facioscapulohumeral muscular dystrophy (FSHD)

Applications: These primary adherent cells are grown in culture to model DUX4 myopathy. Suitable for in vitro testing of drug candidates and mechanistic studies.

Product format: Frozen

Description: Primary fibroblasts isolated from the skeletal muscle of FSHD mouse model described in Dandapat, A. et al. (2014). Cell Rep. 8(5):1484-96.

Phase of Development

Cell lines have been established and characterized.

Desired Partnerships

This technology is now available for:

- License
- Sponsored research
- Co-development

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Researchers

- [Michael Kyba, PhD](#) Position, Department of Pediatrics
- [Darko Bosnakovski, DVM, PhD](#) Assistant Professor, Department of Pediatrics