DUX4 Monoclonal Antibody

ORDERING INFORMATION

Catalog No.: 57100 (clone P4H2)
Size: 100ug in PBS, pH 7.4. Purified by Protein G affinity chromatography.

BACKGROUND

Facioscapulohumeral muscular dystrophy (FSHD) is caused by the deletion of a subset of D4Z4 macro-satellite repeats on chromosome 4. Each repeat contains a retrogene encoding the double-homeobox factor DUX4. DUX4 expression is epi-genetically suppressed in differentiated tissues, and the residual DUX4 transcripts are spliced to remove the carboxyterminal domain that has been associated with cell toxicity. In FSHD individuals, the expression of the full-length DUX4 transcript is not completely suppressed in skeletal muscle and possibly other differentiated tissues.

SPECIFICATION SUMMARY

Antigen: C-terminal 76 amino acids of DUX4 with glutathione-s-transferase (gst) tag.
Host Species: Mouse
Antibody Class: IgG1

SPECIFICITY

This antibody recognizes an epitope at the C-terminus of human DUX4. It does not cross-react with DUX4c.

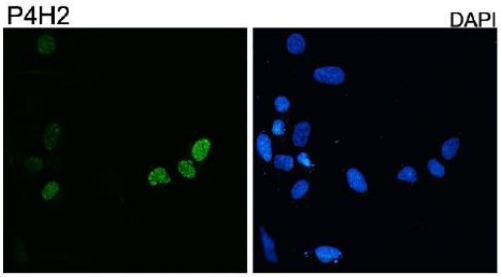
REFERENCE

Snider L et al 2010 PLoS Genetics 6: 1-14.

For in vitro investigational use only. Not intended for diagnostic or therapeutic applications.

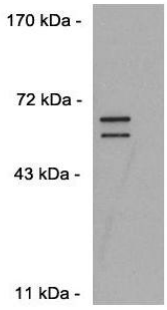
APPLICATIONS

Immunohistochemistry: frozen or paraffin.
Immunofluorescence:



#57100 diluted 1:10,000 on rhabdomyosarcoma cell line RD transfected with pCS2-DUX4. Cells were counterstained with DAPI for nuclei.

Immunoblotting:



#57100 diluted 1:10,000 on C2C12 cells transfected with pCS2-DUX4. There is a doublet because the expression construct contains an upstream alternate translation start codon. The lower band is the canonical size for DUX4.
Positive control: human testis lysate.

See reference below for procedural details. Enduser should determine optimal concentrations for their applications.

DILUTION INSTRUCTIONS

Dilute in PBS or medium which is identical to that used in the assay system.

STORAGE AND STABILITY

This antibody is stable for at least one (1) year at -20°C.

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