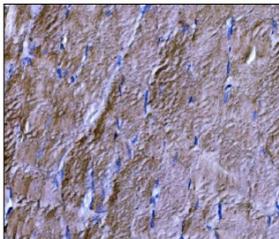


## DYSF Antibody / Dysferlin (RQ6473)

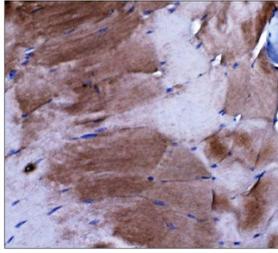
Catalog No.	Formulation	Size
RQ6473	0.5mg/ml if reconstituted with 0.2ml sterile DI water	100 ug

### Bulk quote request

<b>Availability</b>	1-3 business days
<b>Species Reactivity</b>	Human, Mouse, Rat
<b>Format</b>	Antigen affinity purified
<b>Host</b>	Rabbit
<b>Clonality</b>	Polyclonal (rabbit origin)
<b>Isotype</b>	Rabbit IgG
<b>Purity</b>	Affinity purified
<b>Buffer</b>	Lyophilized from 1X PBS with 2% Trehalose
<b>UniProt</b>	O75923
<b>Applications</b>	Western Blot : 1-2ug/ml Immunohistochemistry (FFPE) : 2-5ug/ml Immunofluorescence : 5ug/ml Flow Cytometry : 1-3ug/million cells Direct ELISA : 0.1-0.5ug/ml
<b>Limitations</b>	This DYSF antibody is available for research use only.



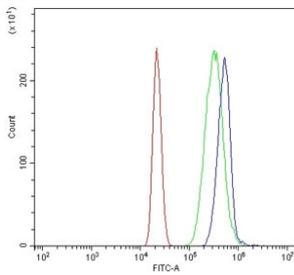
IHC staining of FFPE human skeletal muscle tissue with DYSF antibody. HIER: boil tissue sections in pH8 EDTA for 20 min and allow to cool before testing.



IHC staining of FFPE mouse skeletal muscle tissue with DYSF antibody. HIER: boil tissue sections in pH8 EDTA for 20 min and allow to cool before testing.



Western blot testing of 1) human PC-3, 2) rat heart and 3) mouse heart lysate with DYSF antibody. Predicted molecular weight ~230 kDa.



Flow cytometry testing of human HL60 cells with DYSF antibody at 1ug/million cells (blocked with goat sera); Red=cells alone, Green=isotype control, Blue= DYSF antibody.

## Description

Dysferlin also known as dystrophy-associated fer-1-like protein is a protein that in humans is encoded by the DYSF gene. The protein encoded by this gene belongs to the ferlin family and is a skeletal muscle protein found associated with the sarcolemma. It is involved in muscle contraction and contains C2 domains that play a role in calcium-mediated membrane fusion events, suggesting that it may be involved in membrane regeneration and repair. In addition, the protein encoded by this gene binds caveolin-3, a skeletal muscle membrane protein which is important in the formation of caveolae. Specific mutations in this gene have been shown to cause autosomal recessive limb girdle muscular dystrophy type 2B (LGMD2B) as well as Miyoshi myopathy. Alternative splicing results in multiple transcript variants.

## Application Notes

Optimal dilution of the DYSF antibody should be determined by the researcher.

## Immunogen

An E. coli-derived human protein (amino acids E51-H747) was used as the immunogen for the DYSF antibody.

## Storage

After reconstitution, the DYSF antibody can be stored for up to one month at 4°C. For long-term, aliquot and store at -20°C. Avoid repeated freezing and thawing.

