**BACKGROUND**

Dysferlin is a muscle-specific protein that is essential for normal muscle function and development. Mutations in the human dysferlin gene, DYSF, which maps to chromosome 2p13.3-p13.1, are associated with limb girdle muscular dystrophy-2B (LGMD-2B) and a related, adult-onset, distal dystrophy known as Miyoshi myopathy (MM). Dysferlin, a protein with a molecular mass of approximately 230 kDa, localizes to the muscle fiber membrane, but is absent in MM and LGMD-2B muscle. Dysferlin is detected in 5-6 week embryos, when limbs begin to form regional differentiation. Although it is not essential for initial myogenesis, dysferlin appears to be critical for sustained normal function in mature muscle. It has been suggested that the absence of dysferlin during development gives rise to the disease phenotype in adulthood. Identical mutations in the dysferlin gene can produce more than one myopathy phenotype, indicating that additional genes and/or other factors are also involved in the clinical phenotype. The DYSF gene has no homology to any other known mammalian gene, but the protein product is related to the spermatogenesis factor fer-1 of Caenorhabditis elegans. The name ‘dysferlin’ combines the role of the gene in producing muscular dystrophy with its homology to C. elegans.

**REFERENCES**


**CHROMOSOMAL LOCATION**

Genetic locus: DYSF (human) mapping to 2p13.3; Dysf (mouse) mapping to 6 C3.

**SOURCE**

**PRODUCT**

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin. Blocking peptide available for competition studies, sc-16634 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

**APPLICATIONS**

dysferlin (E-20) is recommended for detection of dysferlin of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for dysferlin siRNA (h): sc-43739, dysferlin siRNA (m): sc-77329, dysferlin shRNA Plasmid (h): sc-43739-SH, dysferlin shRNA Plasmid (m): sc-77329-SH, dysferlin shRNA (h) Lentiviral Particles: sc-43739-V and dysferlin shRNA (m) Lentiviral Particles: sc-77329-V.

**RECOMMENDED SECONDARY REAGENTS**

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000). Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:1000), immunofluorescence: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

**SELECT PRODUCT CITATIONS**


**STORAGE**

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

**RESEARCH USE**

For research use only, not for use in diagnostic procedures.

**PROTOCOLS**

See our web site at www.scbt.com or our catalog for detailed protocols and support products.