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DATA SHEET

Rabbit Anti-Dysferlin Polyclonal: RC0280

Intended Use: For Research Use Only

Description: Dysferlin is the protein product of the 2p13 gene that is defective in patients with Limb-Girdle Muscular Dystrophy type 2B (LGMD2B) and Miyoshi Myopathy (MM). The protein encoded by this gene belongs to the ferlin family and is a skeletal muscle protein found associated with the sarcolemma. It is normally localized to the muscle plasma membrane. In patients with LGMD2B and MM, immunoreactivity to dysferlin is severely reduced or lost, depending on the type of mutation. This antibody is used for the characterization of LGMD2B and MM.

Specifications

Clone:	Polyclonal
Source:	Rabbit
Isotype:	IgG
Reactivity:	Human
Immunogen:	Recombinant human dysferlin protein
Localization:	Membrane
Formulation:	Antibody in PBS pH7.4, containing BSA and ≤ 0.09% sodium azide (NaN3)
Storage:	Store at 2°- 8°C
Applications:	IHC

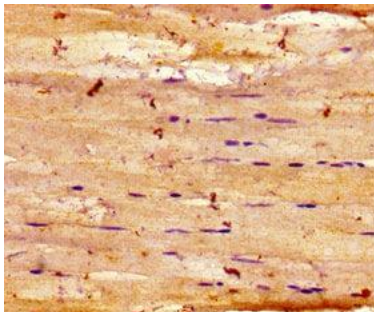
Package:

Description	Catalog No.	Size
Dysferlin Polyclonal Concentrated	RC0280	1 ml

IHC Procedure*

Positive Control Tissue:	Skeletal muscle tissue
Concentrated Dilution:	10-50
Pretreatment:	Citrate pH6.0 or EDTA pH8.0, 15 min Pressure Cooker or 30-60 min water bath at 95°-99°C
Incubation Time and Temp:	Overnight @ 4°C
Detection:	Refer to the detection system manual

* Result should be confirmed by an established diagnostic procedure.



FFPE human skeletal muscle tissue stained with anti-Dysferlin using DAB

References:

1. Dysferlin mediates membrane tubulation and links T-tubule biogenesis to muscular dystrophy. Hofhuis J, et al. J Cell Sci 130:841-852, 2017.
2. DNA-Mediated Gene Therapy in a Mouse Model of Limb Girdle Muscular Dystrophy 2B. Ma J, et al. Mol Ther Methods Clin Dev 7:123-131, 2017.
3. PIK3C2B inhibition improves function and prolongs survival in myotubular myopathy animal models. Sabha N, et al. J Clin Invest 126:3613-25, 2016.
4. A comparison of AAV strategies distinguishes overlapping vectors for efficient systemic delivery of the 6.2?kb Dysferlin coding sequence. Pryadkina M, et al. Mol Ther Methods Clin Dev 2:15009, 2015.

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