Rabbit Anti-Dysferlin Polyclonal: RC0280

Intended Use: For Research Use Only

Description: Emerin is a member of the nuclear lamina associated protein family. It is ubiquitously expressed and localized to the nuclear membrane in normal cells. Mutations of the gene that encodes emerin result in the X-linked recessive disease Emery-Dreifuss muscular dystrophy (EDMD), which is characterized by slowly progressing contractures, skeletal muscle wasting and cardiomyopathy. Reportedly, lack of Emerin expression is one cause of EDMD. Emerin is involved in the association of the nuclear membrane with the lamina, and is localized specifically to desmosomes and fasciae adherents in the heart. Identification of nuclear membrane irregularities with anti-emerin antibody has been reported useful in diagnosing papillary thyroid carcinoma.

Specifications

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

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 minutes using Pressure Cooker, or 30-60 minutes using water bath at 95°-99°C
Incubation Time and Temp:	30-60 minutes @ RT
Detection:	Refer to the detection system manual

* Result should be confirmed by an established diagnostic procedure.

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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