emerin (G-10): sc-398067



The Power to Question

BACKGROUND

Emerin is believed to be 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-Dreyfuss muscular dystrophy (EDMD), which is characterized by slowly progressing contractures, skeletal muscle wasting and cardiomyopathy. Research has demonstrated that the 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 adherentes in the heart. This may account for conduction defects in patients with EDMD.

REFERENCES

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- Bione, S., Small, K., Aksmanovic, V.M., D'Urso, M., Ciccodicola, A., Merlini, L., Morandi, L., Kress, W., Yates, J.R., Warren, S.T. and Toniolo, D. 1995. Identification of new mutations in the Emery-Dreifuss muscular dystrophy gene and evidence for genetic heterogeneity of the disease. Hum. Mol. Genet. 4: 1859-1863.
- Cartegni, L., di Barletta, M.R., Barresi, R., Squarzoni, S., Sabatelli, P., Maraldi, N., Mora, M., Di Blasi, C., Cornelio, F., Merlini, L., Villa, A., Cobianchi, F. and Toniolo, D. 1997. Heart-specific localization of emerin: new insights into Emery-Dreifuss muscular dystrophy. Hum. Mol. Genet. 6: 2257-2264.
- Kubo, S., Tsukahara, T. and Arahata, K. 1997. Emery-Dreifuss muscular dystrophy. Nippon Rinsho 55: 3186-3189.
- Small, K. and Warren, S.T. 1998. Emerin deletions occurring on both Xq28 inversion backgrounds. Hum. Mol. Genet. 7: 135-139.

CHROMOSOMAL LOCATION

Genetic locus: EMD (human) mapping to Xq28.

SOURCE

emerin (G-10) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 186-223 near the C-terminus of emerin of human origin.

PRODUCT

Each vial contains 200 $\mu g \ lgG_{2b}$ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-398067 P, (100 μ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

STORAGE

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

APPLICATIONS

emerin (G-10) is recommended for detection of emerin of human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g of total protein (1 ml of cell lysate)], 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 emerin siRNA (h): sc-35296, emerin shRNA Plasmid (h): sc-35296-SH and emerin shRNA (h) Lentiviral Particles: sc-35296-V.

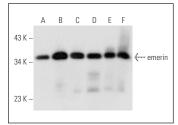
Molecular Weight of emerin: 37 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200, HeLa nuclear extract: sc-2120 or Jurkat nuclear extract: sc-2132.

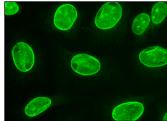
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-lgG κ BP-HRP: sc-516102 or m-lgG κ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz MarkerTM Molecular Weight Standards: sc-2035, UltraCruz[®] Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-lgG κ BP-FITC: sc-516140 or m-lgG κ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz[®] Mounting Medium: sc-24941 or UltraCruz[®] Hard-set Mounting Medium: sc-359850.

DATA







emerin (G-10): sc-398067. Immunofluorescence staining of methanol-fixed HeLa cells showing nuclear envelope localization.

RESEARCH USE

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

PROTOCOLS

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