emerin (MANEM5): sc-81553

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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-Dreifuss 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

1. Bione, S., Maestrini, E., Rivella, S., Mancini, M., Regis, S., Romeo, G. and Toniolo, D. 1994. Identification of a novel X-linked gene responsible for Emery-Dreifuss muscular dystrophy. Nat. Genet. 8: 323-327.

2. Bione, S., Small, K., Aksmanovic, V.M., D'Urso, M., Ciccodicola, A., Merlini, L., Morandi, L., Kress, W., Yates, J.R., Warren, S.T., et al. 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.

3. 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.

4. Kubo, S., Tsukahara, T. and Arahata, K. 1997. Emery-Dreifuss muscular dystrophy. Nippon Rinsho 55: 3186-3189.

5. 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; Emd (mouse) mapping to X A7.3.

SOURCE

emerin (MANEM5) is a mouse monoclonal antibody raised against full length recombinant emerin of human origin.

PRODUCT

Each vial contains 200 µg IgG$_1$ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

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.

APPLICATIONS

emerin (MANEM5) is recommended for detection of emerin of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)] and immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500).

Suitable for use as control antibody for emerin siRNA (h): sc-35296, emerin siRNA (m): sc-35297, emerin siRNA (r): sc-270504, emerin shRNA Plasmid (h): sc-35296-SH, emerin shRNA Plasmid (m): sc-35297-SH, emerin shRNA Plasmid (r): sc-270504-SH, emerin shRNA (h) Lentiviral Particles: sc-35296-V, emerin shRNA (m) Lentiviral Particles: sc-35297-V and emerin shRNA (r) Lentiviral Particles: sc-270504-V.

Molecular Weight of emerin: 34 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200, Saos-2 cell lysate: sc-2235 or K-562 whole cell lysate: sc-2203.

RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ 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-IgGκ BP-FITC: sc-516140 or m-IgGκ 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
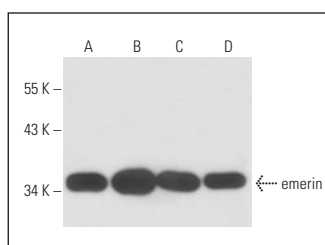
emerin (MANEM5): sc-81553. Western blot analysis of emerin expression in Saos-2 (**A**), K-562 (**B**), HeLa (**C**) and A549 (**D**) whole cell lysates.
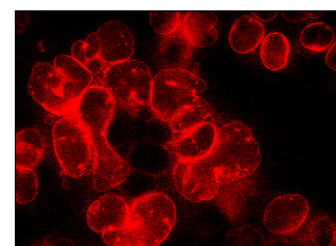
emerin (MANEM5): sc-81553. Immunofluorescence staining of methanol-fixed HeLa cells showing nuclear envelope localization.

PROTOCOLS

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