

emerin (F-11): sc-398278

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. 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.
3. Cartegni, L., di Barletta, M.R., Barresi, R., Squarzone, S., Sabatelli, P., Maraldi, N., Mora, M., Di Blasi, C., Cornelio, F., Merlini, L., Villa, A., Cobiainchi, 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.

SOURCE

emerin (F-11) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 196-223 near the C-terminus of emerin of human origin.

PRODUCT

Each vial contains 200 µg IgG₁ 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-398278 P, (100 µg 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 (F-11) 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 µg per 100-500 µg 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.

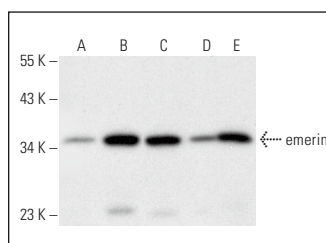
Molecular Weight of emerin: 37 kDa.

Positive Controls: K-562 whole cell lysate: sc-2203, HeLa whole cell lysate: sc-2200 or Jurkat nuclear extract: sc-2132.

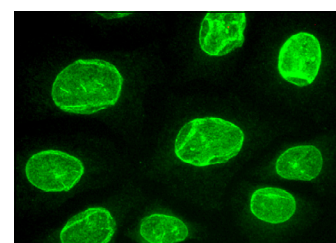
RECOMMENDED SUPPORT REAGENTS

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 (F-11): sc-398278. Western blot analysis of emerin expression in Jurkat (A), K-562 (B) and HeLa (C) whole cell lysates and HeLa (D) and Jurkat (E) nuclear extracts.



emerin (F-11): sc-398278. 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.