BACKGROUND

Newly synthesized Rab proteins are bound to Rab escort proteins (REP) and presented to the Rab geranylgeranyltransferase (GGTase) type II, which mediates the prenylation of Rab proteins on two carboxyterminal cysteine residues. Rab GGTase only recognizes Rab proteins as a substrate when they are bound to REP. REP remains complexed with Rab until it is transported to the appropriate subcellular membrane, although it is still unclear whether REP participates in this targeting. Two isoforms of the REP gene have been isolated, REP-1 and REP-2. The REP-1 gene, located on chromosome Xq21, is prone to a wide variety of mutations, including nonsense, frameshift and splice-site mutations and deletions. In patients with choroideremia (CHM), mutations in the REP-1 gene result in progressive dystrophy of the choroid, retinal pigment epithelium and retina. CHM is an X-linked hereditary eye disease that leads to blindness later in life. REP-2 is able to bind to several Rab proteins with the same affinity as REP-1 and may act as a substitute for REP-1 to prevent widespread tissue abnormalities in patients with CHM.

REFERENCES


CHROMOSOMAL LOCATION

Genetic locus: CHML (human) mapping to 1q43; Chml (mouse) mapping to 1H4.

SOURCE

REP-2 (E-4) is a mouse monoclonal antibody raised against amino acids 58-97 mapping near the N-terminus of REP-2 of mouse origin.

PRODUCT

Each vial contains 200 µg IgG2a kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

REP-2 (E-4) is available conjugated to agarose (sc-398605 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-1807), REP-2 shRNA Plasmid (h): sc-1806-SH, REP-2 shRNA Plasmid (m): sc-1807-SH, REP-2 siRNA (h) Lentiviral Particles: sc-1806-V and REP-2 shRNA (m) Lentiviral Particles: sc-1807-V.

Molecular Weight (predicted) of REP-2: 74 kDa.

Molecular Weight (observed) of REP-2: 90 kDa.

Positive Controls: Neuro-2A whole cell lysate: sc-364185, NIH/3T3 whole cell lysate: sc-2210 or RAW 264.7 whole cell lysate: sc-2211 or RAW 264.7 whole cell lysate: sc-2211.

APPLICATIONS

REP-2 (E-4) is recommended for detection of REP-2 of mouse, rat and 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).


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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: 1:1000-1:1000), 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).

DATA

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.

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