SANTA CRUZ BIOTECHNOLOGY, INC.

REP-1 (2F1): sc-23905

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

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 carboxy-terminal 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 is it 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 Xq21.2, is prone to a wide variety of mutations, including nonsense, frameshift and splice-site mutations and deletions. In patients with choroideraemia (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 a substitute for REP-1 to prevent widespread tissue abnormalities in patients with CHM.

CHROMOSOMAL LOCATION

Genetic locus: CHM (human) mapping to Xq21.2.

SOURCE

REP-1 (2F1) is a mouse monoclonal antibody raised against recombinant human Rab Escort Protein-1 (REP-1).

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.

REP-1 (2F1) is available conjugated to agarose (sc-23905 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-23905 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-23905 PE), fluorescein (sc-23905 FITC), Alexa Fluor® 488 (sc-23905 AF488), Alexa Fluor® 546 (sc-23905 AF546), Alexa Fluor® 594 (sc-23905 AF594) or Alexa Fluor® 647 (sc-23905 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-23905 AF680) or Alexa Fluor® 790 (sc-23905 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

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APPLICATIONS

REP-1 (2F1) is recommended for detection of REP-1 of human origin by Western Blotting (starting dilution 1:500, dilution range 1:500-1:2000) and 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 immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500).

Suitable for use as control antibody for REP-1 siRNA (h): sc-41804, REP-1 shRNA Plasmid (h): sc-41804-SH and REP-1 shRNA (h) Lentiviral Particles: sc-41804-V.

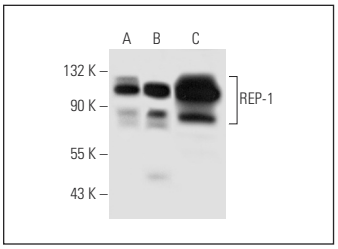
Molecular Weight of REP-1: 83 kDa.

Positive Controls: REP-1 (h): 293T Lysate: sc-117225, A-431 whole cell lysate: sc-2201 or Y79 cell lysate: sc-2240.

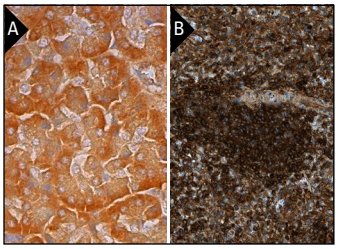
STORAGE

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

DATA



REP-1 (2F1): sc-23905. Western blot analysis of REP-1 expression in non-transfected 293T: sc-117752 (**A**), human REP-1 transfected 293T: sc-117225 (**B**) and Y79 (**C**) whole cell lysates.



REP-1 (2F1): sc-23905. Immunoperoxidase staining of formalin fixed, paraffin-embedded human pancreas tissue showing cytoplasmic staining of exocrine glandular cells (**A**). Immunoperoxidase staining of formalin fixed, paraffin-embedded human spleen tissue showing cytoplasmic staining of cells in red and white pulp high magnification. Kindly provided by The Swedish Human Protein Atlas (HPA) program (**B**).

SELECT PRODUCT CITATIONS

1. Hildebrand, M.S., et al. 2007. Molecular characterization of a novel X-linked syndrome involving developmental delay and deafness. Am. J. Med. Genet. A 143A: 2564-2575.
2. Strunnikova, N.V., et al. 2009. Loss-of-function mutations in Rab escort protein 1 (REP-1) affect intracellular transport in fibroblasts and monocytes of choroideremia patients. PLoS ONE 4: e8402.
3. Esposito, G., et al. 2011. Comprehensive mutation analysis (20 families) of the choroideremia gene reveals a missense variant that prevents the binding of REP1 with Rab geranylgeranyl transferase. Hum. Mutat. 32: 1460-1469.
4. Zhou, Q., et al. 2012. Genetic and phenotypic characteristics of three Mainland Chinese families with choroideremia. Mol. Vis. 18: 309-316.
5. Vasireddy, V., et al. 2013. AAV-mediated gene therapy for choroideremia: preclinical studies in personalized models. PLoS ONE 8: e61396.
6. Furgoch, M.J., et al. 2014. Molecular genetic diagnostic techniques in choroideremia. Mol. Vis. 20: 535-544.
7. Song, K.H., et al. 2017. REP1 inhibits FOXO3-mediated apoptosis to promote cancer cell survival. Cell Death Dis. 8: e2536.
8. Bruiners, N., et al. 2020. The anti-tubercular activity of simvastatin is mediated by cholesterol-driven autophagy via the AMPK-mTORC1-TFEB axis. J. Lipid Res. 61: 1617-1628.
9. Iwagawa, T., et al. 2022. Evaluation of CRISPR/Cas9 exon-skipping vector for choroideremia using human induced pluripotent stem cell-derived RPE. J. Gene Med. E-published.

RESEARCH USE

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