SANTA CRUZ BIOTECHNOLOGY, INC.

EXT2 (A-2): sc-514092

BACKGROUND

Hereditary multiple exostoses (HME) is an autosomal dominant disorder characterized by the formation of exostoses (EXT), which are cartilage-capped bony protuberances mainly located on long bones. Two proteins associated with EXT, EXT1 and EXT2, form homo/heteromeric complexes *in vivo*, which leads to the accumulation of both proteins in the Golgi apparatus. EXT1 and EXT2 are endoplasmic reticulum-localized type II transmembrane glycoproteins that possess, or are tightly associated with, glycosyltransferase activities involved in the polymerization of the glycosaminoglycan, heparan sulfate (HS). EXT2 is a protein that harbors the D-glucuronyl (GlcA) and N-acetyl-D-glucosaminyl (GlcNAc) transferase activities required for biosynthesis of HS. EXT1 rescues defective HS biosynthesis and elevates low GlcA and GlcNAc transferase levels in mutated cells.

CHROMOSOMAL LOCATION

Genetic locus: EXT2 (human) mapping to 11p11.2; Ext2 (mouse) mapping to 2 E1.

SOURCE

EXT2 (A-2) is a mouse monoclonal antibody raised against amino acids 402-537 mapping within an internal region of EXT2 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.

EXT2 (A-2) is available conjugated to agarose (sc-514092 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-514092 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-514092 PE), fluorescein (sc-514092 FITC), Alexa Fluor® 488 (sc-514092 AF488), Alexa Fluor® 546 (sc-514092 AF546), Alexa Fluor® 594 (sc-514092 AF594) or Alexa Fluor® 647 (sc-514092 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-514092 AF680) or Alexa Fluor® 790 (sc-514092 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

APPLICATIONS

EXT2 (A-2) is recommended for detection of EXT2 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).

Suitable for use as control antibody for EXT2 siRNA (h): sc-106830, EXT2 siRNA (m): sc-144985, EXT2 shRNA Plasmid (h): sc-106830-SH, EXT2 shRNA Plasmid (m): sc-144985-SH, EXT2 shRNA (h) Lentiviral Particles: sc-106830-V and EXT2 shRNA (m) Lentiviral Particles: sc-144985-V.

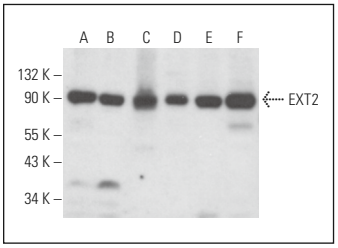
Molecular Weight of EXT2: 90 kDa.

Positive Controls: EXT2 (m): 293T Lysate: sc-126818, MDA-MB-468 cell lysate: sc-2282 or HeLa whole cell lysate: sc-2200.

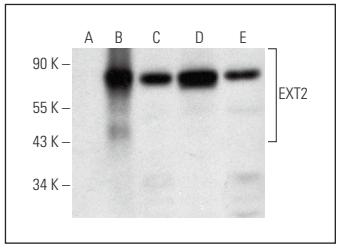
STORAGE

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

DATA



EXT2 (A-2): sc-514092. Western blot analysis of EXT2 expression in A549 (**A**), HeLa (**B**), SCC-4 (**C**), EOC 20 (**D**), 3T3-L1 (**E**) and C6 (**F**) whole cell lysates.



EXT2 (A-2): sc-514092. Western blot analysis of EXT2 expression in non-transfected 293T: sc-117752 (**A**), mouse EXT2 transfected 293T: sc-126818 (**B**), HeLa (**C**), MDA-MB-468 (**D**) and Jurkat (**E**) whole cell lysates.

SELECT PRODUCT CITATIONS

1. Wu, H., et al. 2017. Bone size and quality regulation: concerted actions of mTOR in mesenchymal stromal cells and osteoclasts. Stem Cell Reports 8: 1600-1616.
2. Poli, M., et al. 2019. Hepatic heparan sulfate is a master regulator of hepcidin expression and iron homeostasis in human hepatocytes and mice. J. Biol. Chem. 294: 13292-13303.
3. Puangmalai, N., et al. 2020. Internalization mechanisms of brain-derived Tau oligomers from patients with Alzheimer's disease, progressive supranuclear palsy and dementia with Lewy bodies. Cell Death Dis. 11: 314.
4. Costanzo, M., et al. 2020. Proteomics reveals that methylmalonyl-CoA mutase modulates cell architecture and increases susceptibility to stress. Int. J. Mol. Sci. 21: 4998.
5. Wu, D., et al. 2021. Exostosin1 as a novel prognostic and predictive biomarker for squamous cell lung carcinoma: a study based on bioinformatics analysis. Cancer Med. 10: 2787-2801.
6. Suzuki, T., et al. 2022. Genome-wide CRISPR screen for HSV-1 host factors reveals PAPSS1 contributes to heparan sulfate synthesis. Commun. Biol. 5: 694.
7. Ahat, E., et al. 2022. GRASP depletion-mediated Golgi fragmentation impairs glycosaminoglycan synthesis, sulfation, and secretion. Cell. Mol. Life Sci. 79: 199.
8. Marvian, A.T., et al. 2024. Distinct regulation of Tau Monomer and aggregate uptake and intracellular accumulation in human neurons. Mol. Neurodegener. 19: 100.

RESEARCH USE

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