

EXT1 (A-7): sc-515144

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: EXT1 (human) mapping to 8q24.11; Ext1 (mouse) mapping to 15 C.

SOURCE

EXT1 (A-7) is a mouse monoclonal antibody raised against amino acids 219-332 mapping within an internal region of EXT1 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.

EXT1 (A-7) is available conjugated to agarose (sc-515144 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-515144 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-515144 PE), fluorescein (sc-515144 FITC), Alexa Fluor® 488 (sc-515144 AF488), Alexa Fluor® 546 (sc-515144 AF546), Alexa Fluor® 594 (sc-515144 AF594) or Alexa Fluor® 647 (sc-515144 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-515144 AF680) or Alexa Fluor® 790 (sc-515144 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

Alexa Fluor® is a trademark of Molecular Probes, Inc., Oregon, USA

APPLICATIONS

EXT1 (A-7) is recommended for detection of EXT1 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 EXT1 siRNA (h): sc-106792, EXT1 siRNA (m): sc-144984, EXT1 shRNA Plasmid (h): sc-106792-SH, EXT1 shRNA Plasmid (m): sc-144984-SH, EXT1 shRNA (h) Lentiviral Particles: sc-106792-V and EXT1 shRNA (m) Lentiviral Particles: sc-144984-V.

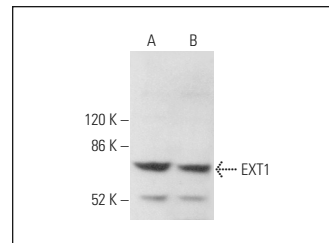
Molecular Weight of EXT1: 86 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200, Jurkat whole cell lysate: sc-2204 or MCF7 whole cell lysate: sc-2206.

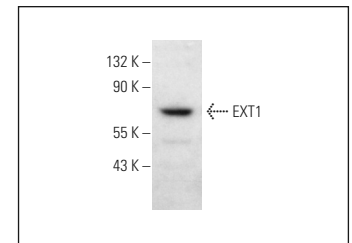
RECOMMENDED SUPPORT REAGENTS

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



EXT1 (A-7): sc-515144. Western blot analysis of EXT1 expression in Jurkat (A) and MCF7 (B) whole cell lysates.



EXT1 (A-7): sc-515144. Western blot analysis of EXT1 expression in HeLa whole cell lysates.

SELECT PRODUCT CITATIONS

- 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.
- 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.
- 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.
- Wilson, L.F.L., et al. 2022. The structure of EXTL3 helps to explain the different roles of bi-domain exostosins in heparan sulfate synthesis. *Nat. Commun.* 13: 3314.
- Dieter, S.M., et al. 2022. Suppression of heparan sulfation re-sensitizes YAP1-driven melanoma to MAPK pathway inhibitors. *Oncogene* 41: 3953-3968.

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.