SANTA CRUZ BIOTECHNOLOGY, INC.

EXT1 (H-114): sc-292430

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

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.

REFERENCES

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2. McCormick, C., Leduc, Y., Martindale, D., Mattison, K., Esford, L. E., Dyer, A.P. and Tufaro, F. 1998. The putative tumour suppressor EXT1 alters the expression of cell-surface heparan sulfate. Nat. Genet. 19: 158-161.
3. Wuyts, W. and Van Hul, W. 2000. Molecular basis of multiple exostoses: mutations in the EXT1 and EXT2 genes. Hum. Mutat. 15: 220-2277.
4. Kobayashi, S., Morimoto, K., Shimizu, T., Takahashi, M., Kurosawa, H. and Shirasawa, T. 2000. Association of EXT1 and EXT2, hereditary multiple exostoses gene products, in Golgi apparatus. Biochem. Biophys. Res. Commun. 268: 860-867.
5. McCormick, C., Duncan, G., Goutsos, K.T. and Tufaro, F. 2000. The putative tumor suppressors EXT1 and EXT2 form a stable complex that accumulates in the Golgi apparatus and catalyzes the synthesis of heparan sulfate. Proc. Natl. Acad. Sci. USA 97: 668-673.

CHROMOSOMAL LOCATION

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

SOURCE

EXT1 (H-114) is a rabbit polyclonal antibody raised against amino acids 219-332 mapping within an internal region of EXT1 of human origin.

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

STORAGE

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

APPLICATIONS

EXT1 (H-114) is recommended for detection of EXT1 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, 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).

EXT1 (H-114) is also recommended for detection of EXT1 in additional species, including equine, canine, bovine, porcine and avian.

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.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 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 goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

RESEARCH USE

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

PROTOCOLS

See our web site at www.scbt.com or our catalog for detailed protocols and support products.