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EXT1

Reactivity: Human Mouse Rat

Tested applications:WB IHC IF

Recommended Dilution:WB 1:500 - 1:2000 IHC 1:50 - 1:200 IF 1:10 - 1:100

Calculated MW:86kDa

Observed MW: Refer to Figures

Immunogen:

Recombinant protein of human EXT1

Storage Buffer:

Store at -20. Avoid freeze / thaw cycles. Buffer: PBS with 0.02% sodium azide, 50% glycerol, pH7.3.

Synonym:

EXT; LGS; TTV; LGCR; TRPS2;

Catalog #:A2030

Antibody Type:

Polyclonal Antibody

Species: Rabbit

Gene ID:2131

Isotype: IgG

Swiss Prot:Q16394

Purity: Affinity purification

For research use only.

Background:

Hereditary multiple exostoses (EXT) is an autosomal dominant disorder characterized by the formation of cartilage-capped tumors (exostoses) that develop from the growth plate of endochondral bone. This condition can lead to skeletal abnormalities, short stature and malignant transformation of exostoses to chondrosarcomas or osteosarcomas. Linkage analyses have identified three different genes for EXT, EXT1 on 8q24.1, EXT2 on 11p11-13 and EXT3 on 19p, a family of tumor suppressor genes. Most EXT cases have been attributed to missense or frameshift mutations, which lead to loss of function of the EXT genes. EXT1 is an ER-resident type II transmembrane glycoprotein and a heparan sulphate polymerase with both D-glucuronyl and N-acetyl-D-glucosaminoglycan transferase activities. Expression of EXT1 in cells results in the alteration of the synthesis and display of cell surface heparan sulfate glycosaminoglycans. EXT1 mutations have been identified in multiple types of human tumors.

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