

# **EXT1** polyclonal antibody

Catalog: BS6597

Host: Ra

Rabbit

Reactivity: Human, Mouse

# **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.

# **Product:**

Rabbit IgG, 1mg/ml in PBS with 0.02% sodium azide, 50% glycerol, pH7.2

**Molecular Weight:** 

# ~ 86 kDa

**Swiss-Prot:** 

#### Q16394

**Purification&Purity:** 

The antibody was affinity-purified from rabbit antiserum by affinity-chromatography using epitope-specific immunogen and the purity is > 95% (by SDS-PAGE).

# **Applications:**

WB 1:500 - 1:2000 IF 1:10 - 1:100

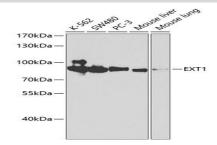
### **Storage&Stability:**

Store at  $4 \,^{\circ}{\rm C}$  short term. Aliquot and store at  $-20 \,^{\circ}{\rm C}$  long term. Avoid freeze-thaw cycles.

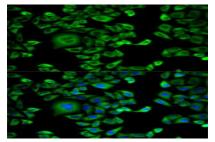
#### **Specificity:**

EXT1 polyclonal antibody detects endogenous levels of EXT1 protein.

#### **DATA:**



Western blot analysis of extracts of various cell lines, using EXT1 antibody at 1:1000 dilution.



Immunofluorescence analysis of HeLa cells using EXT1 antibody. Blue: DAPI for nuclear staining.

#### Note:

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

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