

Rabbit Anti-Exostosin-1/EXT1 Polyclonal: RC0007

Intended Use: For Research Use Only

Description: 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.

Specifications:

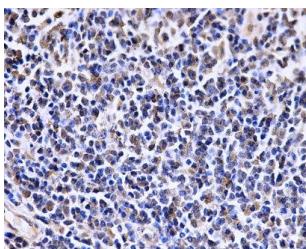
Clone: Polyclonal
Source: Rabbit
Isotype: IgG
Reactivity: Human, mouse, rat
Localization: Endoplasmic reticulum membrane. Golgi apparatus membrane. The EXT1/EXT2 complex is localized in the Golgi apparatus
Formulation: Antibody in PBS pH7.4, containing BSA, and $\leq 0.09\%$ sodium azide (NaN₃)
Storage: Store at 2°- 8°C
Applications: IHC, WB
Package:

Description	Catalog No.	Size
Exostosin-1/EXT1 Polyclonal Concentrated	RC0007	1 ml

IHC Procedure*:

Positive Control Tissue: Tonsilrectum tissue
Concentrated Dilution: 10-50
Pretreatment: Citrate pH6.0 or EDTA pH8.0, 15 minutes using Pressure Cooker, or 30-60 minutes using water bath at 95°-99°C
Incubation Time and Temp: 30-60 minutes @ RT
Detection: Refer to the detection system manual

* Result should be confirmed by an established diagnostic procedure.



FFPE human esophageal cancer stained with anti-EXT1 using DAB

References:

1. Syndecans and Enzymes Involved in Heparan Sulfate Biosynthesis and Degradation Are Differentially Expressed During Human Odontogenesis. Kero D, et al. Front Physiol 9:732, 2018.
2. Novel EXT1 mutation identified in a pedigree with hereditary multiple exostoses. Cao L, et al. Oncol Rep. Feb;31(2):713-8, 2014.
3. Exome sequencing and functional analysis identifies a novel mutation in EXT1 gene that causes multiple osteochondromas. Zhang F, et al. PLoS One 8:e72316, 2013.

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Rev. A