Rabbit Anti-Exostosin-2/EXT2 Polyclonal: RC0168

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
Localization:	Endoplasmic reticulum membrane, Golgi apparatus membrane
Formulation:	Antibody in PBS pH7.4, containing BSA, and ≤ 0.09% sodium azide (NaN3)
Storage:	Store at 2°- 8°C
Applications:	IHC, ELISA

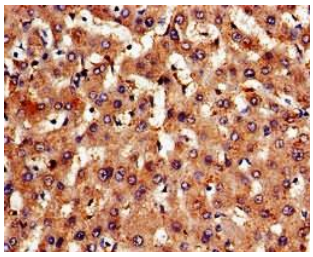
Package:

Description	Catalog No.	Size
Exostosin-2/EXT2 Polyclonal Concentrated	RC0168	1 ml

IHC Procedure*:

Positive Control Tissue:	Colon, liver
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:	Overnight @ 4°C
Detection:	Refer to the detection system manual

* Result should be confirmed by an established diagnostic procedure.



FFPE human liver stained with anti-EXT2 using DAB

References:

1. Mutational Analysis of Exostosin 1 and 2 Genes in Multiple Osteochondroma. Malini K et al. Indian J Pediatr. Jul;82(7):649-50, 2015.
2. Reduced Expression of EXTL2, a Member of the Exostosin (EXT) Family of Glycosyltransferases, in Human Embryonic Kidney 293 Cells Results in Longer Heparan Sulfate Chains. Katta K, et al. J Biol Chem. May 22;290(21):13168-77, 2015.
3. A splice mutation and mRNA decay of EXT2 provoke hereditary multiple exostoses. Tian C, et al. PLoS One 9:e94848, 2014.

Orders: customercare@medaysis.com Support: techsupport@medaysis.com Tel: 510-509-3153 www.medaysis.com