abcam

Product datasheet

Recombinant Human Ext2 protein ab158396

1 Image

Overview

Product name	Recombinant Human Ext2 protein
Protein length	Protein fragment

Description

Nature	Recombinant
Source	Wheat germ
Amino Acid Sequence	
Species	Human
Sequence	GFSTWTYRQGYDVSIPVYSPLSAEVDLPEKGPGPRQY FLLSSQVGLHPEY REDLEALQVKHGESVLVLDKCTNLSEGVLSVRKRCHK HQVFDYPQVLQEA
Amino acids	216 to 315
Tags	GST tag N-Terminus

Specifications

Our Abpromise guarantee covers the use of **ab158396** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Applications	ELISA Western blot
Form	Liquid
Additional notes	Protein concentration is above or equal to 0.05 mg/ml.

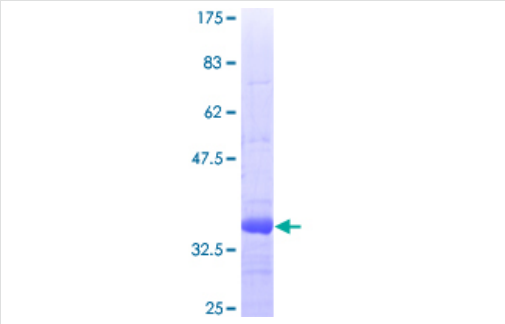
Preparation and Storage

Stability and Storage	Shipped on dry ice. Upon delivery aliquot and store at -80ºC. Avoid freeze / thaw cycles. pH: 8.00 Constituents: 0.31% Glutathione, 0.79% Tris HCl

General Info

Function	Glycosyltransferase required for the biosynthesis of heparan-sulfate. The EXT1/EXT2 complex possesses substantially higher glycosyltransferase activity than EXT1 or EXT2 alone. Appears to be a tumor suppressor.
Tissue specificity	Ubiquitous.
Pathway	Protein modification; protein glycosylation.
Involvement in disease	Defects in EXT2 are a cause of hereditary multiple exostoses type 2 (EXT2) [MIM:133701]. EXT is a genetically heterogeneous bone disorder caused by genes segregating on human chromosomes 8, 11, and 19 and designated EXT1, EXT2 and EXT3 respectively. EXT is a dominantly inherited skeletal disorder primarily affecting endochondral bone during growth. The disease is characterized by formation of numerous cartilage-capped, benign bone tumors (osteocartilaginous exostoses or osteochondromas) that are often accompanied by skeletal deformities and short stature. In a small percentage of cases exostoses have exhibited malignant transformation resulting in an osteosarcoma or chondrosarcoma. Osteochondromas development can also occur as a sporadic event. Defects in EXT2 are the cause of Potocki-Shaffer syndrome (PSS) [MIM:601224]. It is a contiguous gene syndrome due to proximal deletion of chromosome 11p11.2, including EXT2 and ALX4.
Sequence similarities	Belongs to the glycosyltransferase 47 family.
Cellular localization	Endoplasmic reticulum membrane. Golgi apparatus membrane. The EXT1/EXT2 complex is localized in the Golgi apparatus.

Images



SDS-PAGE - Recombinant Human Ext2 protein (ab158396)

ab158396 on a 12.5% SDS-PAGE stained with Coomassie Blue.

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