

## Product datasheet

# Anti-Ext1 antibody ab126305

★★★★★ [1 Abreviews](#) [6 References](#) [4 Images](#)

### Overview

---

<b>Product name</b>	Anti-Ext1 antibody
<b>Description</b>	Rabbit polyclonal to Ext1
<b>Host species</b>	Rabbit
<b>Tested applications</b>	<b>Suitable for:</b> IHC-P
<b>Species reactivity</b>	<b>Reacts with:</b> Human
<b>Immunogen</b>	antigen sequence, corresponding to amino acids 205-335 of Human Ext1.
<b>Positive control</b>	IHC-P: Human rectum, placenta, liver and prostate tissue.
<b>General notes</b>	<p>The Life Science industry has been in the grips of a reproducibility crisis for a number of years. Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets your needs before purchasing.</p> <p>If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be found below, along with publications, customer reviews and Q&amp;As</p>

### Properties

---

<b>Form</b>	Liquid
<b>Storage instructions</b>	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid repeated freeze / thaw cycles.
<b>Storage buffer</b>	pH: 7.20 Preservative: 0.02% Sodium azide Constituents: 40% Glycerol (glycerin, glycerine), 59% PBS
<b>Purity</b>	Immunogen affinity purified
<b>Clonality</b>	Polyclonal
<b>Isotype</b>	IgG

### Applications

---

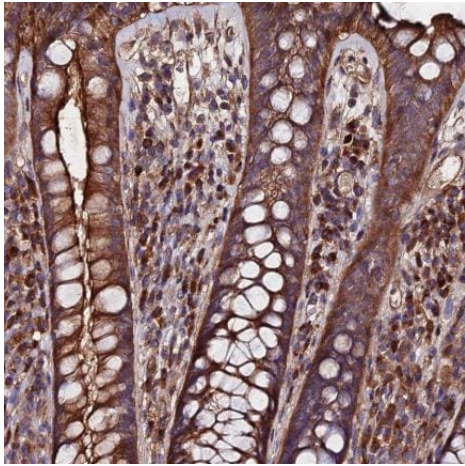
**The Abpromise guarantee** Our **Abpromise guarantee** covers the use of ab126305 in the following tested applications. The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Application	Abreviews	Notes
IHC-P	★★★★★ (1)	1/50 - 1/200. Perform heat mediated antigen retrieval with citrate buffer pH 6 before commencing with IHC staining protocol.

## Target

<b>Function</b>	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.
<b>Tissue specificity</b>	Ubiquitous.
<b>Pathway</b>	Protein modification; protein glycosylation.
<b>Involvement in disease</b>	<p>Defects in EXT1 are a cause of hereditary multiple exostoses type 1 (EXT1) [MIM:133700]. 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 (osteochondrogenous 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.</p> <p>Defects in EXT1 are a cause of tricho-rhino-phalangeal syndrome type 2 (TRPS2) [MIM:150230]. A syndrome that combines the clinical features of trichorhinophalangeal syndrome type 1 and multiple exostoses type 1. Affected individuals manifest multiple dysmorphic facial features including large, laterally protruding ears, a bulbous nose, an elongated upper lip, as well as sparse scalp hair, winged scapulae, multiple cartilaginous exostoses, redundant skin, and mental retardation. Note=A chromosomal aberration resulting in the loss of functional copies of TRPS1 and EXT1 has been found in TRPS2 patients.</p> <p>Defects in EXT1 are a cause of chondrosarcoma (CHDSA) [MIM:215300]. It is a malignant neoplasm derived from cartilage cells. Chondrosarcomas range from slow-growing non-metastasizing lesions to highly aggressive metastasizing sarcomas.</p>
<b>Sequence similarities</b>	Belongs to the glycosyltransferase 47 family.
<b>Cellular localization</b>	Endoplasmic reticulum membrane. Golgi apparatus membrane. The EXT1/EXT2 complex is localized in the Golgi apparatus.

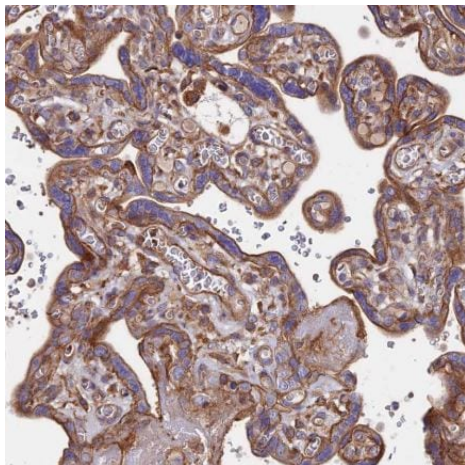
## Images



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Ext1 antibody (ab126305)

Immunohistochemical analysis of human rectum tissue labeling Ext1 in the cytoplasm of glandular cells with ab126305 at a 1/50 dilution.

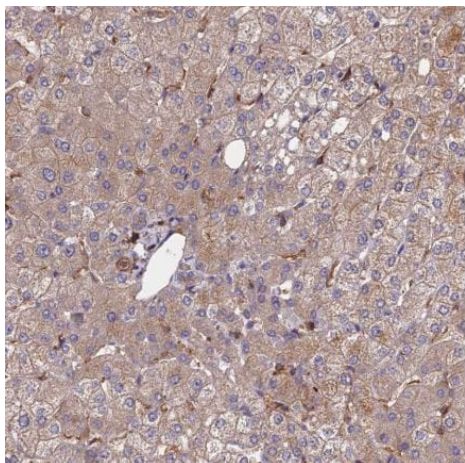
Performed heat mediated antigen retrieval with citrate buffer pH 6 before commencing with IHC staining protocol.



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Ext1 antibody (ab126305)

Immunohistochemical analysis of human placenta tissue labeling Ext1 in the cytoplasm of trophoblastic cells with ab126305 at a 1/50 dilution.

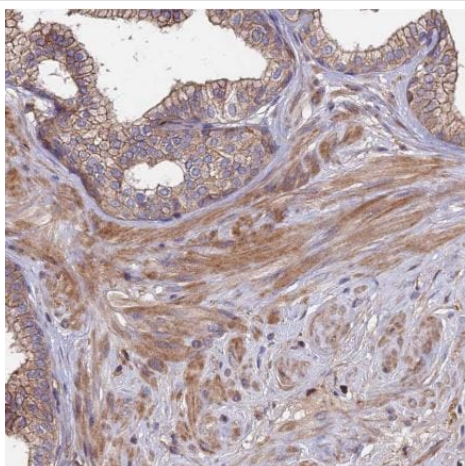
Performed heat mediated antigen retrieval with citrate buffer pH 6 before commencing with IHC staining protocol.



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Ext1 antibody (ab126305)

Immunohistochemical analysis of human liver tissue labeling Ext1 in the cytoplasm of hepatocytes with ab126305 at a 1/50 dilution.

Performed heat mediated antigen retrieval with citrate buffer pH 6 before commencing with IHC staining protocol.



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Ext1 antibody (ab126305)

Immunohistochemical analysis of human prostate tissue labeling Ext1 in the cytoplasm of smooth muscle cells with ab126305 at a 1/50 dilution.

Performed heat mediated antigen retrieval with citrate buffer pH 6 before commencing with IHC staining protocol.

**Please note:** All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

#### **Our Abpromise to you: Quality guaranteed and expert technical support**

- Replacement or refund for products not performing as stated on the datasheet
- Valid for 12 months from date of delivery
- Response to your inquiry within 24 hours

- We provide support in Chinese, English, French, German, Japanese and Spanish
- Extensive multi-media technical resources to help you
- We investigate all quality concerns to ensure our products perform to the highest standards

If the product does not perform as described on this datasheet, we will offer a refund or replacement. For full details of the Abpromise, please visit <https://www.abcam.com/abpromise> or contact our technical team.

### **Terms and conditions**

---

- Guarantee only valid for products bought direct from Abcam or one of our authorized distributors