# abcam

### Product datasheet

# Alexa Fluor® 488 Anti-Sonic Hedgehog antibody [EP1190Y] ab203961



1 References 2 图像

#### 概述

产品名称 Alexa Fluor® 488荧光Anti-Sonic Hedgehog抗体[EP1190Y]

描述 Alexa Fluor® 488荧光兔单克隆抗体[EP1190Y] to Sonic Hedgehog

**宿主** Rabbit

偶联物 Alexa Fluor® 488. Ex: 495nm, Em: 519nm

 经测试应用
 适用于: ICC/IF

 种属反应性
 与反应: Human

免疫原 Synthetic peptide. This information is proprietary to Abcam and/or its suppliers.

阳性对照 ICC/IF: HepG2 cells.

常规说明 This product is a recombinant monoclonal antibody, which offers several advantages including:

- High batch-to-batch consistency and reproducibility
- Improved sensitivity and specificity
- Long-term security of supply
- Animal-free production

For more information see here.

Our RabMAb<sup>®</sup> technology is a patented hybridoma-based technology for making rabbit monoclonal antibodies. For details on our patents, please refer to **RabMAb**<sup>®</sup> **patents**.

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#### 性能

形式 Liquid

**存放说明** Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C.

Stable for 12 months at -20°C. Store In the Dark.

**存储溶液** pH: 7.40

Preservative: 0.02% Sodium azide

Constituents: 30% Glycerol (glycerin, glycerine), 1% BSA, PBS

纯**度** Protein A purified

同种型 lgG

#### 应用

#### The Abpromise guarantee

Abpromise™承诺保证使用ab203961于以下的经测试应用

"应用说明"部分 下显示的仅为推荐的起始稀释度;实际最佳的稀释度/浓度应由使用者检定。

应用	Ab评论	说明
ICC/IF		1/200. This product gave a positive signal in HepG2 cells fixed with 4% formaldehyde (10 min) and 100% methanol (5 min).

#### 靶标

#### 功能

Binds to the patched (PTC) receptor, which functions in association with smoothened (SMO), to activate the transcription of target genes. In the absence of SHH, PTC represses the constitutive signaling activity of SMO. Also regulates another target, the gli oncogene. Intercellular signal essential for a variety of patterning events during development: signal produced by the notochord that induces ventral cell fate in the neural tube and somites, and the polarizing signal for patterning of the anterior-posterior axis of the developing limb bud. Displays both floor plate- and motor neuron-inducing activity. The threshold concentration of N-product required for motor neuron induction is 5-fold lower than that required for floor plate induction.

组织特异性

 $\label{thm:expressed} \textbf{Expressed in fetal intestine, liver, lung, and kidney. Not expressed in adult tissues.}$ 

疾病相关

Defects in SHH are the cause of microphthalmia isolated with coloboma type 5 (MCOPCB5) [MIM:611638]. Microphthalmia is a clinically heterogeneous disorder of eye formation, ranging from small size of a single eye to complete bilateral absence of ocular tissues. Ocular abnormalities like opacities of the cornea and lens, scaring of the retina and choroid, cataract and other abnormalities like cataract may also be present. Ocular colobomas are a set of malformations resulting from abnormal morphogenesis of the optic cup and stalk, and the fusion of the fetal fissure (optic fissure).

Defects in SHH are the cause of holoprosencephaly type 3 (HPE3) [MIM:142945]. Holoprosencephaly (HPE) [MIM:236100] is the most common structural anomaly of the brain, in which the developing forebrain fails to correctly separate into right and left hemispheres.

Holoprosencephaly is genetically heterogeneous and associated with several distinct facies and phenotypic variability. The majority of HPE3 cases are apparently sporadic, although clear examples of autosomal dominant inheritance have been described. Interestingly, up to 30% of obligate carriers of HPE3 gene in autosomal dominant pedigrees are clinically unaffected. Defects in SHH are a cause of solitary median maxillary central incisor (SMMCI) [MIM:147250]. SMMCI is a rare dental anomaly characterized by the congenital absence of one maxillary central incisor.

Defects in SHH are the cause of triphalangeal thumb-polysyndactyly syndrome (TPTPS) [MIM:174500]. TPTPS is an autosomal dominant syndrome characterized by a wide spectrum of pre- and post-axial abnormalities due to altered SHH expression pattern during limb development. TPTPS mutations have been mapped to the 7q36 locus in the LMBR1 gene which contains in its intron 5 a long-range cis-regulatory element of SHH expression.

Belongs to the hedgehog family.

The C-terminal domain displays an autoproteolysis activity and a cholesterol transferase activity. Both activities result in the cleavage of the full-length protein and covalent attachment of a cholesterol moiety to the C-terminal of the newly generated N-terminal fragment (N-product). The N-product is the active species in both local and long-range signaling, whereas the C-product has no signaling activity.

Cholesterylation is required for N-product targeting to lipid rafts and multimerization. N-palmitoylation of Cys-24 by HHAT is required for N-product multimerization and full activity.

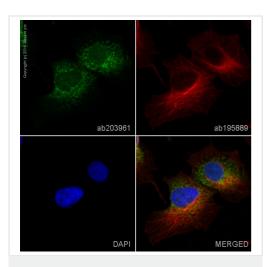
Cell membrane. The N-product either remains associated with lipid rafts at the cell surface, or forms freely diffusible active multimers with its hydrophobic lipid-modified N- and C-termini buried inside and Secreted > extracellular space. The C-terminal peptide diffuses from the cell.

序列相似性

翻译后修饰

细胞定位

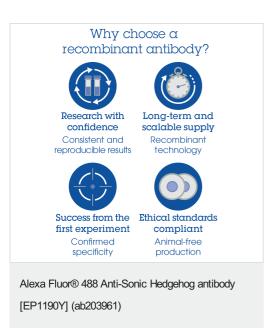
## 图片



Immunocytochemistry/ Immunofluorescence - Alexa Fluor® 488 Anti-Sonic Hedgehog antibody [EP1190Y] (ab203961) ab203961 staining Sonic Hedgehog in HepG2 cells. The cells were fixed with 4% formaldehyde (10 min), permeabilized with 0.1% Triton X-100 for 5 minutes and then blocked with 1% BSA/10% normal goat serum/0.3M glycine in 0.1% PBS-Tween for 1h. The cells were then incubated overnight at +4°C with ab203961 at 1/200 dilution (shown in green) and <a href="mailto:ab195889">ab195889</a>, Mouse monoclonal to alpha Tubulin (Alexa Fluor® 594), at 1/250 dilution (shown in red). Nuclear DNA was labelled with DAPI (shown in blue).

Image was taken with a confocal microscope (Leica-Microsystems, TCS SP8).

This product also gave a positive signal under the same testing conditions in HepG2 cells fixed with 100% methanol (5min).



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