

Recombinant mouse Sonic Hedgehog protein ab75431

1 图像

描述	
产品名称	重组小鼠Sonic Hedgehog蛋白
生物活性	Determined by its ability to induce alkaline phosphatase production by C3H10T1/2 fibroblasts, with an expected ED50 in the range of 0.05 - 0.25 ug/ml.
纯度	> 95 % SDS-PAGE. Purity is greater than 97% as determined by reducing and non-reducing SDS-PAGE and analytical HPLC.
表达系统	Escherichia coli
蛋白长度	Protein fragment
无动物成分	No
性质	Recombinant
种属	Mouse
序列	MIIGPGRGFG KRRHPKKLTP LAYKQFIPNV AEKTLGASGR YEGKITRNSE RFKELTPNYN PDIIFKDEEN TGADRLMTQR CKDKLNALAI SVMNQWPGVK LRVTEGWDED GHHSEESLHY EGRAVDITTS DRDRSKYGML ARLAVEAGFD WVYYESKAHI HCSVKAENSV AAKSGG
氨基酸	26 to 198
技术指标	
Our Abpromise guarantee covers the use of ab75431 in the following tested applications.	
The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.	
应用	SDS-PAGE Functional Studies
形式	Lyophilized
制备和贮存	
稳定性和存储	Shipped at 4°C. Upon delivery aliquot. Store at -80°C. Avoid freeze / thaw cycle. Constituent: 0.164% Sodium phosphate

This product is an active protein and may elicit a biological response in vivo, handle with caution.

复溶

Reconstitute with sterile water at a concentration of 0.1 - 0.5 mg/ml, which can be further diluted into other aqueous solutions.

常规信息

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

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, scarring 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.

序列相似性

Belongs to the hedgehog family.

翻译后修饰

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.

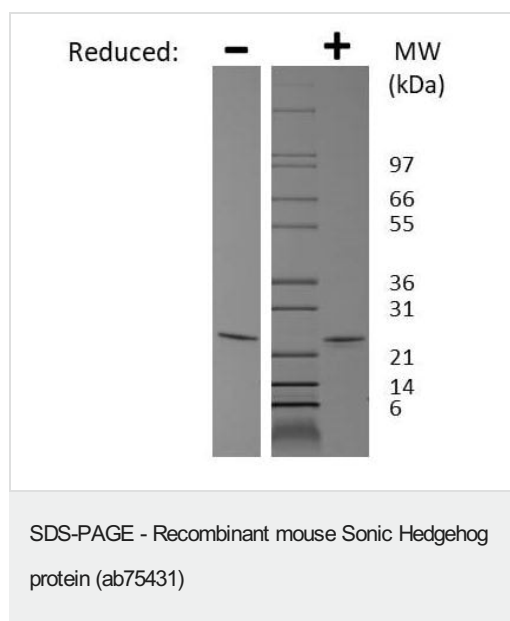
Cholesterylolation is required for N-product targeting to lipid rafts and multimerization.

N-palmitoylation of Cys-24 by HHAT is required for N-product multimerization and full activity.

细胞定位

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.

图片



SDS PAGE analysis of ab75431 under non-reducing (-) and reducing (+) conditions. Stained with Coomassie Blue.

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