

Recombinant human Sonic Hedgehog protein (Active) ab82127

描述	
产品名称	重组人Sonic Hedgehog蛋白(Active)
生物活性	Biological Activity : Determined by its ability to induce alkaline phosphatase production by C3H/10T1/2 (CCL-226) cells. The expected ED ₅₀ for this effect is 0.8-1.0 µg/ml.
纯度	> 98 % SDS-PAGE.
内毒素水平	< 1.000 Eu/µg
表达系统	Escherichia coli
Accession	<u>Q15465</u>
蛋白长度	Full length protein
无动物成分	No
性质	Recombinant
种属	Human
序列	IVIGPGRGFGKRRHPKKLTPLAYKQFIPNVAEKTLGASGRYE GKISRNSE RFKELTPNYNPDIIFKDEENTGADRLMTQRCKDKLNALAI SV MNQWPGVK LRVTEGWDEDGHHSEESLHYEGRALDITTSRDRSKYGLAR LAVEAGFD WVYESKAHIHCSVKAENSVAAKSGG
预测分子量	20 kDa
氨基酸	22 to 197
额外的序列信息	176 amino acid residues, including an N-terminal Ile-Val-Ile sequence substituted for the naturally occurring, chemically modified, Cys residue. Corresponding to Sonic Hedgehog protein N-product.

技术指标	
Our <u>Abpromise guarantee</u> covers the use of ab82127 in the following tested applications.	
The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.	
应用	Functional Studies SDS-PAGE
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.

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

复溶

Reconstitute in water to a concentration of 0.1- 1.0 mg/ml. Do not vortex. This solution can be stored at 2-8degC for up to 1 week. For extended storage, it is recommended to further dilute in a buffer containing a carrier protein (example 0.1% BSA) and store in working aliquots at -20 to -80degC.

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.

细胞定位

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.

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.cn/abpromise> or contact our technical team.

Terms and conditions

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