abcam

Product datasheet

Recombinant human Sonic Hedgehog protein (Active) ab82127

製品の詳細

製品名	Recombinant human Sonic Hedgehog protein (Active)
生理活性	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.
精製度	> 98 % SDS-PAGE.
エンドトキシン・レベル	< 1.000 Eu/µg
発現系	Escherichia coli
アクセッション番号	Q15465
タンパク質長	Full length protein
Animal free	No
由来	Recombinant
生物種	Human
配列	IVIGPGRGFGKRRHPKKLTPLAYKQFIPNVAEKTLGASGRYEGKISRNSE RFKELTPNYNPDIIFKDEENTGADRLMTQRCKDKLNALAISVMNQWPGVK LRVTEGWDEDGHHSEESLHYEGRALDITTSDRDRSKYGMLARLAVEAGFD WVYYESKAHIHCSVKAENSVAAKSGG
予測される分子量	20 kDa
領域	22 to 197
配列の追加情報	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.

特性

Our **Abpromise guarantee** 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.

アプリケーション	Functional Studies
	SDS-PAGE
製品の状態	Lyophilized

前処理および保存

保存方法および安定性

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.

再構成

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.

関連情報

機能

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.

組織特異性

Expressed in fetal intestine, liver, lung, and kidney. Not expressed in adult tissues.

関連疾患

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.

配列類似性

Belongs to the hedgehog family.

翻訳後修飾

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.
細胞内局在	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.

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