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Product datasheet

Recombinant Human Sonic Hedgehog protein ab174054

1 Image

Description

Product name	Recombinant Human Sonic Hedgehog protein
Purity	> 95 % SDS-PAGE.
Endotoxin level	< 1.000 Eu/µg
Expression system	HEK 293 cells
Accession	**Q15465**
Protein length	Protein fragment
Animal free	No
Nature	Recombinant
Species	Human
Sequence	CGPGRGFGKRRHPKKLTPLAYKQFIPNVAEKTLGASGRYE GKISRNSERF KELTPNYNPD IIFKDEENTGADRLMTQRCKDKLNALAISVMNQWPGVK LRVTEGWDEDGHHSEESLHYEG RAVDITTSDRDRSKYGMLARLAVEAG FDWVYYESKAHIHCSVKAENSVAAKSGG
Predicted molecular weight	20 kDa including tags
Amino acids	24 to 197
Tags	His tag C-Terminus
Additional sequence information	Sequence corresponds to the cleaved Sonic hedgehog protein N-product chain. (Accession # NP_000184).

Specifications

Our **Abpromise guarantee** covers the use of **ab174054** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Applications	SDS-PAGE
Form	Lyophilized

Preparation and Storage

Stability and Storage

Shipped at 4°C. Store at 4°C prior to reconstitution. Store at -20°C or -80°C. Avoid freeze / thaw cycle. Reconstitute for long term storage.

pH: 7.40
Constituents: 5% Trehalose, 95% PBS

Reconstitution

It is recommended to reconstitute the lyophilized product in 50 µl sterile deionized water to a final concentration of 1 mg/ml. Solubilize for 30 to 60 minutes at room temperature with occasional gentle mixing. Carrier protein (0.1% HSA or BSA) is strongly recommended for further dilution and long term storage.

General Info

Function

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.

Tissue specificity

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

Involvement in disease

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.

Sequence similarities

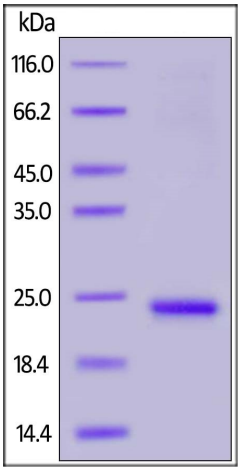
Belongs to the hedgehog family.

Post-translational modifications

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.
Cellular localization	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.

Images



SDS-PAGE - Recombinant Human Sonic Hedgehog protein (ab174054)

SDS-PAGE of reduced ab174054 stained overnight with Coomassie Blue. The protein migrates as 23-25 kDa due to glycosylation.

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