# abcam

# Product datasheet

# Anti-Sonic Hedgehog antibody [RM0128-4A37] ab86462

# ★★★★★ 1 Abreviews 2 References

#### 製品の概要

製品名 Anti-Sonic Hedgehog antibody [RM0128-4A37]

製品の詳細 Rat monoclonal [RM0128-4A37] to Sonic Hedgehog

**由来種** Rat

特異性 ab86462 detects Sonic Hedgehog, but not dhh or ihh.

アプリケーション 適用あり: IHC-P, WB

種交差性 交差種: Mouse

免疫原 Recombinant fragment from the C terminal region of mouse Sonic Hedgehog

特記事項

ab86462 was produced from a hybridoma (mouse myeloma fused with spleen cells from an

immunized rat).

The Life Science industry has been in the grips of a reproducibility crisis for a number of years. Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets

your needs before purchasing.

If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be

found below, along with publications, customer reviews and Q&As

#### 製品の特性

製品の状態 Liquid

保存方法 Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid repeated freeze / thaw cycles.

バッファー Constituent: PBS

精製度 Immunogen affinity purified

特記事項(精製) lgG fraction of culture supernatant purified by Protein A/G affinity chromatography and 0.2 μm

filtered.

一次抗体 備考 ab86462 was produced from a hybridoma (mouse myeloma fused with spleen cells from an

immunized rat).

**ポリ/モノ** モノクローナル **ウローン名** RM0128-4A37

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#### アプリケーション

**The Abpromise guarantee** <u>Abpromise保証は、</u>次のテスト済みアプリケーションにおけるab86462の使用に適用されます アプリケーションノートには、推奨の開始希釈率がありますが、適切な希釈率につきましてはご検討ください。

アプリケーション	Abreviews	特記事項
IHC-P	<b>★★★★</b> <u>(1)</u>	Use at an assay dependent concentration.
WB		Use at an assay dependent concentration.

### ターゲット情報

#### 機能

# 組織特異性

関連疾患

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.

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

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.

#### 配列類似性

#### 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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