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DHH Rabbit pAb

Catalog # AP97488

Product Information

Application	WB, IHC-P, IHC-F, IF, E
Primary Accession	O43323
Other Accession	O43323
Predicted	Rat, Pig, Human, Mouse, Dog, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	43577
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human DHH
Epitope Specificity	101-200/396
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	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). This covalent modification appears to play an essential role in restricting the spatial distribution of the protein activity to the cell surface. The N-product is the active species in both local and long-range signaling, whereas the C-product has no signaling activity (By similarity).
DISEASE	Defects in DHH may be the cause of partial gonadal dysgenesis with minifascicular neuropathy 46,XY (PGD) [MIM:607080]. PGD is characterized by the presence of a testis on one side and a streak or an absent gonad at the other, persistence of Muellerian duct structures, and a variable degree of genital ambiguity.Defects in DHH may be the cause of complete pure gonadal dysgenesis 46,XY type (GDXYM) [MIM:233420]; also known as male-limited gonadal dysgenesis 46,XY. GDXYM is a type of hypogonadism in which no functional gonads are present to induce puberty in an externally female person whose karyotype is then found to be XY. The gonads are found to be non-functional streaks.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	This gene encodes a member of the Hedgehog family. The hedgehog gene family encodes signaling molecules that play an important role in regulating morphogenesis. This protein is predicted to be made as a precursor that is autocatalytically cleaved; the N terminal portion is soluble and contains the signalling activity while the C terminal portion is involved in precursor processing. More importantly, the C terminal product covalently attaches a cholesterol moiety to the N terminal product, restricting the N terminal product to the cell surface and preventing it from freely diffusing throughout the organism. Defects in this protein have been associated with partial gonadal dysgenesis (PGD) accompanied by minifascicular polyneuropathy. This protein may be involved in both male gonadal differentiation and

perineurial development.

Additional Information

Gene ID	50846
Other Names	GDMN; GDXYM; HHG-3; SRXY7; DHH_HUMAN; DHH; 3.1.-.-; DHH_MOUSE; desert hedgehog signaling molecule; desert hedgehog (Drosophila) homolog; desert hedgehog
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500,ELISA=1:5000-10000
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Protein Information

Name	DHH {ECO:0000303|PubMed:33063110, ECO:0000312|HGNC:HGNC:2865}
Function	[Desert hedgehog protein]: Precursor of desert hedgehog, a morphogen that activates the smoothened signaling pathway, and which is essential for a variety of patterning events during development (By similarity). The C-terminal part of the precursor displays an autoproteolysis and a cholesterol transferase activity, resulting (1) in the cleavage of the full-length protein into two parts, desert hedgehog protein N-product and C-product and (2) covalent attachment of a cholesterol moiety to the C-terminus of the newly generated N-product (DHH-N) (By similarity). Both autoproteolysis and a cholesterol transferase activities occur in the endoplasmic reticulum (By similarity). Following additional lipidation, DHH-N acts as a morphogen, while the C-product is degraded in the endoplasmic reticulum (By similarity).
Cellular Location	[Desert hedgehog protein]: Endoplasmic reticulum membrane {ECO:0000250|UniProtKB:Q15465}. Golgi apparatus membrane {ECO:0000250|UniProtKB:Q15465}. Note=Co-localizes with HHAT in the ER and Golgi membrane. {ECO:0000250|UniProtKB:Q15465}

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