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SUFU Rabbit pAb

Catalog # AP99324

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q9UMX1
Other Accession	Q9UMX1
Reactivity	Rat, Mouse
Predicted	Pig, Human, Chicken, Dog, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	53947
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human SUFU
Epitope Specificity	433-484/484
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 SUFU family.
SUBUNIT	May form homodimers. Part of a DNA-bound corepressor complex containing SAP18, GLI1 and SIN3. Part of a complex containing CTNNB1. Binds BTRC, GLI2, GLI3, SAP18 and STK36. Binds both free and DNA-bound GLI1. Interacts with KIF7. Interacts with GLI3FL and this interaction regulates the formation of either repressor or activator forms of GLI3. Its association with GLI3FL is regulated by Hh signaling and dissociation of the SUFU-GLI3 interaction requires the presence of the ciliary motor KIF3A. Interacts with ULK3; inactivating the protein kinase activity of ULK3.
DISEASE	Defects in SUFU are a cause of medulloblastoma (MDB) [MIM:155255]. MDB is a malignant, invasive embryonal tumor of the cerebellum with a preferential manifestation in children. Defects in SUFU play a role in predisposition to desmoplastic MDB. These tumors make up about 20 to 30% of medulloblastomas, have a more nodular architecture than 'classical' medulloblastoma, and may have a better prognosis.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	SUFU is a key negative regulator in the vertebrate Hedgehog signaling pathway. SUFU interacts with genes encoding proteins in this signal transduction pathway. In Drosophila, Intracellular transduction of the Hedgehog pathway involves the release of a large complex containing SUFU. SUFU inhibits the activity of the transcription factor Gli1 and interacts with Gli2, Gli3 and the serine/threonine kinase Fused. SUFU is widely expressed in adult and embryonic tissues with higher expression in tissues patterned by hegdehog signaling. The SUFU gene locus maps to a region that is deleted in glioblastomas, prostate cancer, malignant melanoma and endometrial cancer.

Additional Information

Gene ID	51684
Other Names	BCNS2; JBTS32; PRO1280; SUFUH; SUFUXL; SUFU_HUMAN; SUFU; SUFU negative regulator of hedgehog signaling; suppressor of fused homolog (Drosophila)
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
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	SUFU {ECO:0000303|PubMed:12068298, ECO:0000312|HGNC:HGNC:16466}
Function	Negative regulator in the hedgehog/smoothened signaling pathway that acts by sequestering the GLI (GI1, GLI2 and GLI3) transcription factors in the cytoplasm (PubMed:10559945, PubMed:10564661, PubMed:10806483, PubMed:12068298, PubMed:12975309, PubMed:15367681, PubMed:22365972, PubMed:24217340, PubMed:24311597, PubMed:27234298, PubMed:28965847, PubMed:35831023). When smoothened signaling is initiated, GLI transcription factors dissociate from SUFU, translocate to the nucleus and activate expression of target genes (PubMed:24311597, PubMed:28965847, PubMed:35831023). In absence of smoothened signaling, the SUFU-GLI3 complex is recruited to cilia, leading to the efficient processing of full-length GLI3 into transcription repressor GLI3R: SUFU participates to GLI3 processing by promoting recruitment of GSK3B to GLI3 (PubMed:24311597, PubMed:28965847). May also act as a negative regulator of beta-catenin signaling (By similarity). Required for normal embryonic development (By similarity). Required for the proper formation of hair follicles and the control of epidermal differentiation (By similarity).
Cellular Location	Cytoplasm. Nucleus. Cell projection, cilium {ECO:0000250|UniProtKB:Q9Z0P7}. Note=Recruited to cilia in response to smoothened signaling, leading to dissociation from GLI (GI1, GLI2 and GLI3) transcription factors. {ECO:0000250|UniProtKB:Q9Z0P7}
Tissue Location	Ubiquitous in adult tissues. Detected in osteoblasts of the perichondrium in the developing limb of 12-week old embryos. Isoform 1 is detected in fetal brain, lung, kidney and testis Isoform 2 is detected in fetal testis, and at much lower levels in fetal brain, lung and kidney.

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