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Gli3 Rabbit pAb

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Catalog # AP54547

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	P10071
Reactivity	Rat, Mouse
Predicted	Human, Rabbit, Chicken, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	169863
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Gli3
Epitope Specificity	481-570/1580
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Nucleus. Cytoplasm. Cell projection
DISEASE	Defects in GLI3 are the cause of Greig cephalo-poly-syndactyly syndrome (GCPS) [MIM:175700]. GCPS is an autosomal dominant disorder affecting limb and craniofacial development. It is characterized by pre- and postaxial polydactyly, syndactyly of fingers and toes, macrocephaly and hypertelorism. Defects in GLI3 are a cause of Pallister-Hall syndrome (PHS) [MIM:146510]. PHS is characterized by a wide range of clinical manifestations. It mainly associates central or postaxial polydactyly, syndactyly, and hypothalamic hamartoma. Malformations are frequent in the viscera, e.g. anal atresia, bifid uvula, congenital heart malformations, pulmonary or renal dysplasia. It is an autosomal dominant disorder. Defects in GLI3 are a cause of type A1/B postaxial polydactyly (PAPA1/PAPB) [MIM:174200, 603596]. PAPA in humans is an autosomal dominant trait characterized by an extra digit in the ulnar and/or fibular side of the upper and/or lower extremities. The extra digit is well formed and articulates with the fifth, or extra, metacarpal/metatarsal, and thus it is usually functional. Defects in GLI3 are a cause of polydactyly preaxial type 4 (POP4) [MIM:174700]. Polydactyly preaxial type 4 (i.e., polydactyly on the radial/tibial side of the hand/foot) covers a heterogeneous group of entities. In preaxial polydactyly type IV, the thumb shows only the mildest degree of duplication, and syndactyly of various degrees affects fingers 3 and 4. Defects in GLI3 are the cause of acrocallosal syndrome (ACS) [MIM:200990]; also abbreviated ACLS. ACS is characterized by postaxial polydactyly, hallux duplication, macrocephaly, and absence of the corpus callosum, usually with severe developmental delay.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	It has long been known that the overexpression of either Wnt-1 or the GLI proteins results in cancer; however, the molecular basis for this transformation was poorly understood. The Wnt-1 and GLI proteins have now been placed in a signaling cascade downstream of the mammalian homologs

of the Drosophila hedgehog and patched proteins. The Drosophila segment polarity gene hedgehog (hh) encodes a secreted protein that appears to function in embryonic and imaginal disc patterning. The ptc gene, also identified as a Drosophila segment polarity gene, encodes the transmembrane protein patched, the expression of which is precisely regulated during embryonic development. Hedgehog has been shown to enhance the expression of the Wnt family of proteins through a signaling cascade involving the GLI transcription factors, while patched functions as a repressor opposing the effects of hedgehog. Mutations in the ptc gene, which result in unregulated hedgehog signaling, have been correlated with the most common type of cancer, basal cell carcinoma, which affects 750,000 individuals annually in the United States alone.

Additional Information

Gene ID	2737
Other Names	Transcriptional activator GLI3, GLI3 form of 190 kDa, GLI3-190, GLI3 full-length protein, GLI3FL, Transcriptional repressor GLI3R, GLI3 C-terminally truncated form, GLI3 form of 83 kDa, GLI3-83, GLI3
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	GLI3 {ECO:0000303|PubMed:2118997, ECO:0000312|HGNC:HGNC:4319}
Function	Transcription factor that acts as a key effector of the smoothened signaling pathway, and which plays a role in the development and patterning of various body structures, including the brain and limbs (PubMed:10075717, PubMed:10375510, PubMed:10693759, PubMed:11238441, PubMed:17764085, PubMed:20154143, PubMed:27146893). Specifically binds to the minimal GLI-consensus sequence 5'-GACCACCCA- 3' to activate or repress the expression of target genes (PubMed:10075717). Functions both as a transcriptional activator and repressor: the full-length GLI3 form (Transcription activator GLI3) acts as a transcription activator following smoothened activation, while the Transcription repressor GLI3R, which is generated in absence of smoothened, acts as a transcription repressor (PubMed:10375510). The repressor form (GLI3R) constitutes the major form of GLI3 (PubMed:10375510).
Cellular Location	Nucleus. Cytoplasm. Cell projection, cilium. Note=Full-length GLI3 is sequestered in the cytoplasm by SUFU in absence of smoothened signaling [Transcription activator GLI3]: Nucleus. Cytoplasm Cell projection, cilium Note=Sequestered in the cytoplasm by SUFU in absence of smoothened signaling (PubMed:38968120). Accumulates in primary cilia following smoothened activation: cilium localization requires the presence of KIF7 (PubMed:19592253, PubMed:20154143). It then translocates to the nucleus, where is activates transcription of target genes (PubMed:38968120). Translocation to the nucleus is promoted by interaction with ZIC1 (PubMed:11238441). TMEM216 reduces its nuclear localization (By similarity). {ECO:0000250|UniProtKB:Q61602, ECO:0000269|PubMed:11238441,

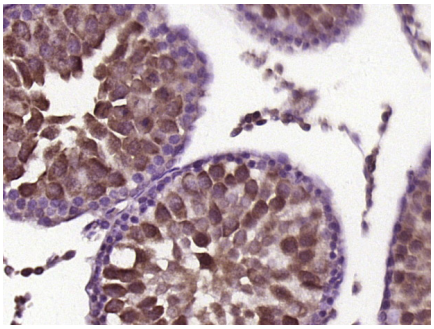
ECO:0000269|PubMed:19592253, ECO:0000269|PubMed:20154143, ECO:0000269|PubMed:38968120}

Tissue Location Is expressed in a wide variety of normal adult tissues, including lung, colon, spleen, placenta, testis, and myometrium.

Background

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

Images



Paraformaldehyde-fixed, paraffin embedded (rat testis tissue); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (Gli3) Polyclonal Antibody, Unconjugated (AP54547) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.