abcam

Product datasheet

Anti-Gli3 antibody ab6050

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Overview

Product name	Anti-Gli3 antibody
Description	Rabbit polyclonal to Gli3
Host species	Rabbit
Tested applications	**Suitable for:** ICC/IF, IHC-Fr, WB, IHC-P
Species reactivity	**Reacts with:** Mouse, Human **Predicted to work with:** Rat, Chicken, Xenopus laevis
Immunogen	Synthetic peptide: NEDESPGQTYHRERRNA-C conjugated to KLH, corresponding to amino acids 41-57 of Human GLI3. Run BLAST with Run BLAST with
General notes	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

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.
Storage buffer	Preservative: 0.01% Sodium azide Constituents: 0.42% Potassium phosphate, 0.87% Sodium chloride
Purity	Immunogen affinity purified
Clonality	Polyclonal
Isotype	IgG

Applications

The Abpromise guarantee Our **Abpromise guarantee** covers the use of ab6050 in the following tested applications.

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

Application	Abreviews	Notes
ICC/IF	★★☆☆☆ (1)	Use a concentration of 1 µg/ml. See Abreview.
IHC-Fr	★★★☆☆ (1)	Use at an assay dependent concentration.
WB		Use at an assay dependent concentration. Predicted molecular weight: 170 kDa. PubMed: 23293081
IHC-P		Use a concentration of 0.5 - 5 µg/ml.

Target

Function Has a dual function as a transcriptional activator and a repressor of the sonic hedgehog (Shh) pathway, and plays a role in limb development. The full-length GLI3 form (GLI3FL) after phosphorylation and nuclear translocation, acts as an activator (GLI3A) while GLI3R, its C-terminally truncated form, acts as a repressor. A proper balance between the GLI3 activator and the repressor GLI3R, rather than the repressor gradient itself or the activator/repressor ratio gradient, specifies limb digit number and identity. In concert with TRPS1, plays a role in regulating the size of the zone of distal chondrocytes, in restricting the zone of PTHLH expression in distal cells and in activating chondrocyte proliferation. Binds to the minimal GLI-consensus sequence 5'-GGGTGGTC-3'.

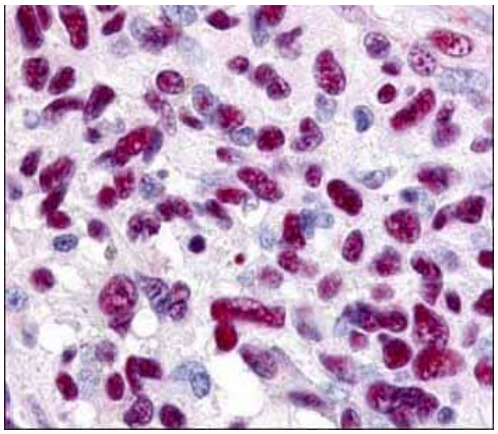
Tissue specificity Is expressed in a wide variety of normal adult tissues, including lung, colon, spleen, placenta, testis, and myometrium.

Involvement in 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.

Sequence similarities Belongs to the GLI C2H2-type zinc-finger protein family.
Contains 5 C2H2-type zinc fingers.

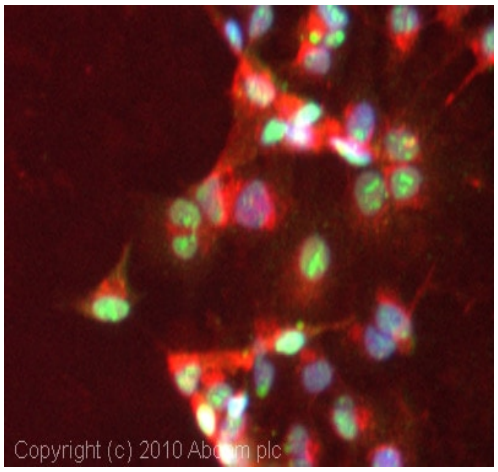
Post-translational modifications	Phosphorylated on multiple sites by protein kinase A (PKA) and phosphorylation by PKA primes further phosphorylation by CK1 and GSK3. Phosphorylation is essential for its proteolytic processing. Transcriptional repressor GLI3R, a C-terminally truncated form, is generated from the full-length GLI3 protein (GLI3FL/GLI3-190) through proteolytic processing. This process requires PKA-primed phosphorylation of GLI3, ubiquitination of GLI3 and the presence of BTRC. GLI3FL is complexed with SUFU in the cytoplasm and is maintained in a neutral state. Without the Hh signal, the SUFU-GLI3 complex is recruited to cilia, leading to the efficient processing of GLI3FL into GLI3R. GLI3R formation leads to its dissociation from SUFU, allowing it to translocate into the nucleus, and repress Hh target genes. When Hh signaling is initiated, SUFU dissociates from GLI3FL and this has two consequences. First, GLI3R production is halted. Second, free GLI3FL translocates to the nucleus, where it is phosphorylated, destabilized, and converted to a transcriptional activator (GLI3A). Phosphorylated in vitro by ULK3.
Cellular localization	Nucleus. Cytoplasm. Cell projection > cilium. GLI3FL is localized predominantly in the cytoplasm while GLI3R resides mainly in the nucleus. Ciliary accumulation requires the presence of KIF7 and SMO. Translocation to the nucleus is promoted by interaction with ZIC1.

Images



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Gli3 antibody (ab6050)

ab6050 at 0.625 µg/ml staining human glioblastoma.



Immunocytochemistry/ Immunofluorescence - Anti-Gli3 antibody (ab6050)

ICC/IF image of ab6050 stained HepG2 cells. The cells were 4% formaldehyde fixed (10 min) and then incubated in 1%BSA / 10% normal goat serum / 0.3M glycine in 0.1% PBS-Tween for 1h to permeabilise the cells and block non-specific protein-protein interactions. The cells were then incubated with the antibody (ab6050, 1µg/ml) overnight at +4°C. The secondary antibody (green) was Alexa Fluor® 488 goat anti-rabbit IgG (H+L) used at a 1/1000 dilution for 1h. Alexa Fluor® 594 WGA was used to label plasma membranes (red) at a 1/200 dilution for 1h. DAPI was used to stain the cell nuclei (blue) at a concentration of 1.43µM.

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