abcam

Product datasheet

Anti-Gli3 antibody [EPR4594] ab181130

Recombinant RabMAb

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Overview

Product name	Anti-Gli3 antibody [EPR4594]
Description	Rabbit monoclonal [EPR4594] to Gli3
Host species	Rabbit
Tested applications	**Suitable for:** IHC-P, ICC/IF, WB
Species reactivity	**Reacts with:** Human **Predicted to work with:** Chimpanzee
Immunogen	Recombinant fragment. This information is proprietary to Abcam and/or its suppliers.
Positive control	WB: 293 lysate ICC/IF: 293 cells and human kidney tissue.
General notes	This product is a recombinant monoclonal antibody, which offers several advantages including: - High batch-to-batch consistency and reproducibility - Improved sensitivity and specificity - Long-term security of supply - Animal-free production For more information **see here**. Our RabMAb® technology is a patented hybridoma-based technology for making rabbit monoclonal antibodies. For details on our patents, please refer to **RabMAb® patents**.

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C long term. Avoid freeze / thaw cycle.
Storage buffer	Preservative: 0.01% Sodium azide Constituents: 40% Glycerol, 0.05% BSA, 59% PBS
Purity	Protein A purified
Clonality	Monoclonal
Clone number	EPR4594
Isotype	IgG

Applications

The Abpromise guarantee Our **Abpromise guarantee** covers the use of ab181130 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
IHC-P	★★★★★ (1)	1/100. Perform heat mediated antigen retrieval with Tris/EDTA buffer pH 9.0 before commencing with IHC staining protocol.
ICC/IF	★★★★☆ (1)	1/100.
WB	★★★★☆ (1)	1/1000 - 1/2000. Predicted molecular weight: 190 kDa.

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'.

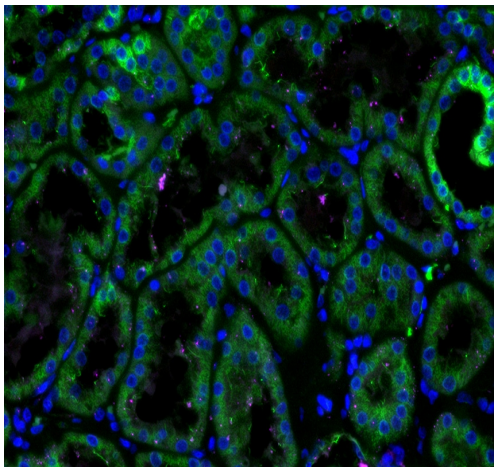
Tissue specificity Is expressed in a wide variety of normal adult tissues, including lung, colon, spleen, placenta, testis, and myometrium.

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.

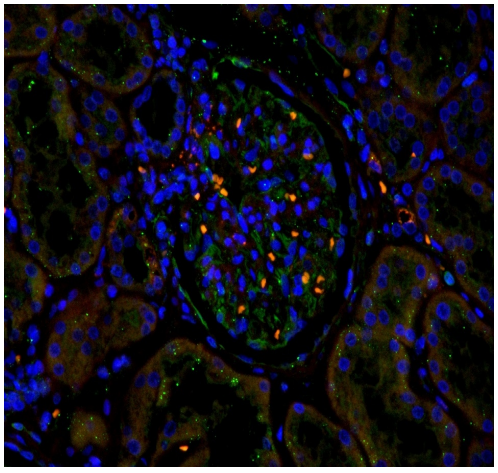
Images



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Gli3 antibody [EPR4594] (ab181130)

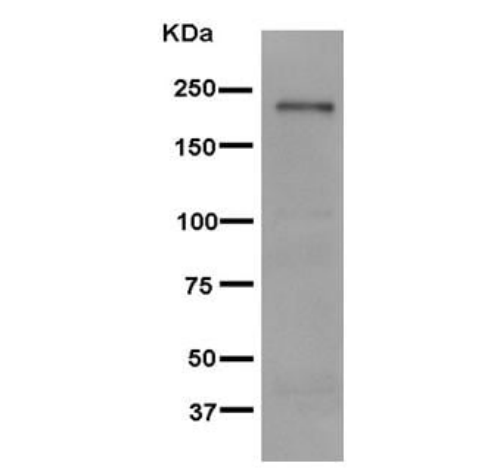
This image is courtesy of an abreview by Lindsey Fitzsimons

Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) analysis of Human kidney tissue labelling Gli3 (green) with ab181130 at 1/100 dilution. Antigen retrieval was performed using Tris/EDTA buffer pH 9 for 30 minutes, a HRP-conjugated goat anti-rabbit IgG (H+L) (Alexa Fluor® 488) **ab150078** was used as the secondary antibody (1/1000). Sections were co-stained with Monoclonal Anti-tubulin (1/1000). Magnification at x100.



Immunocytochemistry/ Immunofluorescence - Anti-Gli3 antibody [EPR4594] (ab181130)

Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) analysis of Human kidney tissue labelling Gli3 (green) with ab181130 at 1/100 dilution. Antigen retrieval was performed using Tris/EDTA buffer pH 9 for 30 minutes, a HRP-conjugated goat anti-rabbit IgG (H+L) (Alexa Fluor® 488) **ab150078** was used as the secondary antibody (1/1000). Sections were co-stained with Monoclonal Anti-tubulin (1/1000). Magnification at x40.



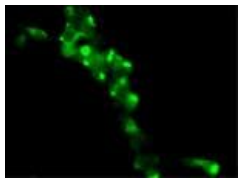
Western blot - Anti-Gli3 antibody [EPR4594] (ab181130)

Anti-Gli3 antibody [EPR4594] (ab181130) at 1/1000 dilution + 293 lysate at 20 µg

Secondary

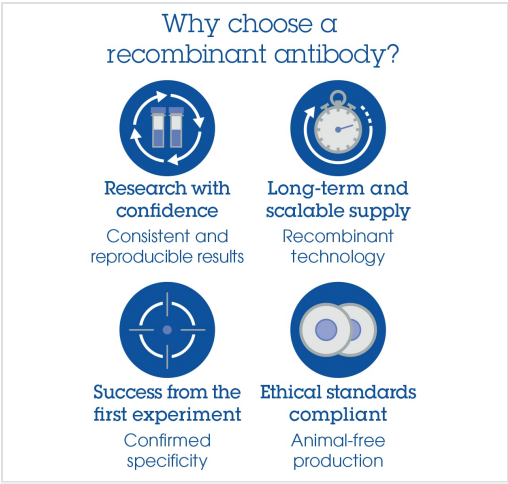
Goat Anti-Rabbit IgG, (H+L), Peroxidase conjugated at 1/1000 dilution

Predicted band size: 190 kDa



Immunocytochemistry/ Immunofluorescence - Anti-Gli3 antibody [EPR4594] (ab181130)

Immunofluorescence analysis of 293 cells (fixative 4% paraformaldehyde) labeling Gli3 with ab181130 at a 1/100 dilution. Goat anti rabbit IgG (Alexa Fluor 488™) secondary used at a 1/200 diution.



Anti-Gli3 antibody [EPR4594] (ab181130)

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