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Product datasheet

Anti-FGFR3 antibody ab10649

3 References 1 Image

Overview

Product name	Anti-FGFR3 antibody
Description	Rabbit polyclonal to FGFR3
Host species	Rabbit
Tested applications	**Suitable for:** WB, IP, IHC-P
Species reactivity	**Reacts with:** Human
Immunogen	Synthetic peptide: KDLLPPAPPSSGGSRT, corresponding to amino acids 792-806 of the cytoplasmic region of Human FGFR3 with N-terminal added lysine. The peptide is conjugated to KLH with glutaraldehyde. Run BLAST with ExPASy Run BLAST with NCBI
Positive control	Whole cell lysates of transfected 293T (embryonic kidney) cells expressing recombinant human FGFR3.
General notes	Fibroblast growth factors (FGFs) are members of a large family of structurally related polypeptides (17-38kD) that are potent physiological regulators of growth and differentiation in a wide variety of cells of mesodermal, ectodermal and endodermal origin. FGFs are substantially involved in normal development, wound healing and repair, angiogenesis, a variety of neurotrophic activities, in hematopoiesis as well as in tissue remodeling and maintenance. They have also been implicated in pathological conditions such as tumorigenesis and metastasis. The FGF family consists of at least seventeen members designated FGF1 through FGF17. To date, four genes encoding for high affinity cell surface FGF receptors (FGFRs) have been identified: FGFR1 [flg1, cek1], FGFR2 [bek, cek3], FGFR3 [cek2] and FGFR4. Soluble, secreted or possibly cleaved forms of FGFR1 and FGFR2 have also been found in body fluids or were artificially constructed. FGFRs are members of the tyrosine kinase family of growth factor receptors. They are glycosylated 110-150 kD proteins that are constructed of an extracellular ligand binding region with either two or typically three immunoglobulin (Ig)-like domains and an eight amino acid 'acidic box', a transmembrane region and a cytoplasmic split tyrosine kinase domain that is activated following ligand binding and receptor dimerization. The ligand binding site of all FGFRs is confined to the extracellular Ig-like domains 2 and 3. FGFRs exhibit overlapping recognition and redundant specificity. One receptor type may bind several of the FGFs with a similar affinity. Also one FGF type may bind similarly to several distinct receptors. This accounts for the rather identical effects of different FGF ligands on common cell types. FGFs binding to cellular FGFRs depends on, or is markedly facilitated by, the low-affinity interaction of FGFs with the polysaccharide component of cell surface or extracellular matrix heparan sulfate proteoglycans (HSPG). For example, perlecan, a basement membrane HSPG, promotes high affinity binding of FGF2 in vitro and angiogenesis in vivo. Signal transduction by FGFRs requires dimerization or oligomerization and autophosphorylation of the receptors through their tyrosine kinase domain.

Subsequent association with cytoplasmic signaling molecules leads to DNA synthesis or differentiation. The signaling and biological responses elicited by distinct FGFRs substantially differ and are dictated by the intracellular domain. FGFR3 is widely expressed in many fetal and adult human and animal tissues. FGFR3 expression profile largely correlates with its tissue specific expression at the mRNA level. It is considered the only FGFR expressed in the Organ of Corti of the rat cochlea. Tissue cultured cells transfected with the full length FGFR3 cDNA display the expected membrane localization of the receptor. Interestingly, nuclear localization (nucleoli excluded) of FGFR3 attributable to a 110kD splice variant, has been reported for normal and breast cancer cells. Deletions of chromosome 4p encompassing the FGFR3 gene cause the Wolf-Hirshhorn syndrome (growth failure, mental retardation, cardiac and bone malformations). Achondroplasia is an inherited disorder in which growth abnormality of bone or cartilage leads to skeletal maldevelopment and dwarfism. It is associated with recurrent mutations of a single amino acid in the transmembrane domain of the FGFR3 protein.

The peptide immunogen is conjugated to KLH with glutaraldehyde.

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: 0.097% Sodium azide Constituents: 0.0268% PBS, 1% BSA
Purity	Immunogen affinity purified
Purification notes	The product is affinity purified on an immunizing peptide-agarose column.
Primary antibody notes	Fibroblast growth factors (FGFs) are members of a large family of structurally related polypeptides (17-38kD) that are potent physiological regulators of growth and differentiation in a wide variety of cells of mesodermal, ectodermal and endodermal origin. FGFs are substantially involved in normal development, wound healing and repair, angiogenesis, a variety of neurotrophic activities, in hematopoiesis as well as in tissue remodeling and maintenance. They have also been implicated in pathological conditions such as tumorigenesis and metastasis. The FGF family consists of at least seventeen members designated FGF1 through FGF17. To date, four genes encoding for high affinity cell surface FGF receptors (FGFRs) have been identified: FGFR1 [flg1, cek1], FGFR2 [bek, cek3], FGFR3 [cek2] and FGFR4. Soluble, secreted or possibly cleaved forms of FGFR1 and FGFR2 have also been found in body fluids or were artificially constructed. FGFRs are members of the tyrosine kinase family of growth factor receptors. They are glycosylated 110-150 kD proteins that are constructed of an extracellular ligand binding region with either two or typically three immunoglobulin (Ig)-like domains and an eight amino acid ‘acidic box’, a transmembrane region and a cytoplasmic split tyrosine kinase domain that is activated following ligand binding and receptor dimerization. The ligand binding site of all FGFRs is confined to the extracellular Ig-like domains 2 and 3. FGFRs exhibit overlapping recognition and redundant specificity. One receptor type may bind several of the FGFs with a similar affinity. Also one FGF type may bind similarly to several distinct receptors. This accounts for the rather identical effects of different FGF ligands on common cell types. FGFs binding to cellular FGFRs depends on, or is markedly facilitated by, the low-affinity interaction of FGFs with the polysaccharide component of cell surface or extracellular matrix heparan sulfate proteoglycans (HSPG). For example, perlecan, a basement membrane HSPG, promotes high affinity binding of FGF2 in vitro and angiogenesis in vivo. Signal transduction by FGFRs requires dimerization or oligomerization and autophosphorylation of the receptors through their tyrosine kinase domain.

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Clonality Polyclonal

Isotype IgG

Applications

The Abpromise guarantee Our **Abpromise guarantee** covers the use of ab10649 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
WB		1/1000. Detects a band of approximately 110-120 kDa. 1/1000 as determined by western blotting using a whole cell extract of transfected 293T cells expressing recombinant human FGFR3.
IP		1/2000. 1/2000 as determined by immunoprecipitation using a whole cell lysate of transfected 293T cells expressing recombinant human FGFR3.
IHC-P		1/1 - 1/250. The epitope(s) recognized by the antibody is resistant to routine formalin-fixation and paraffin embedding.

Target

Function Receptor for acidic and basic fibroblast growth factors. Preferentially binds FGF1.

Tissue specificity Expressed in brain, kidney and testis. Very low or no expression in spleen, heart, and muscle. In 20- to 22-week old fetuses it is expressed at high level in kidney, lung, small intestine and brain, and to a lower degree in spleen, liver, and muscle. Isoform 2 is detected in epithelial cells. Isoform 1 is not detected in epithelial cells. Isoform 1 and isoform 2 are detected in fibroblastic cells.

Involvement in disease Defects in FGFR3 are the cause of achondroplasia (ACH) [MIM:100800]. ACH is an autosomal dominant disease and is the most frequent form of short-limb dwarfism. It is characterized by a long, narrow trunk, short extremities, particularly in the proximal (rhizomelic) segments, a large head with frontal bossing, hypoplasia of the midface and a trident configuration of the hands. Defects in FGFR3 are the cause of Crouzon syndrome with acanthosis nigricans (CAN)

[MIM:612247]. Classic Crouzon disease which is caused by mutations in the FGFR2 gene is characterized by craniosynostosis (premature fusion of the skull sutures), and facial hypoplasia. Crouzon syndrome with acanthosis nigricans (a skin disorder characterized by pigmentation anomalies), CAN, is considered to be an independent disorder from classic Crouzon syndrome. CAN is characterized by additional more severe physical manifestation, such as Chiari malformation, hydrocephalus, and atresia or stenosis of the choanas, and is caused by a specific mutation (Ala-391 to Glu) in the transmembrane domain of FGFR3. It is proposed to have an autosomal dominant mode of inheritance.
Defects in FGFR3 are a cause of thanatophoric dysplasia type (TD) [MIM:187600, 187601]; also known as thanatophoric dwarfism or platyspondylic lethal skeletal dysplasia Sand Diego type (PLSD-SD). TD is the most common neonatal lethal skeletal dysplasia. Affected individuals display features similar to those seen in homozygous achondroplasia. It causes severe shortening of the limbs with macrocephaly, narrow thorax and short ribs. In the most common subtype, TD1, femur are curved, while in TD2, straight femurs are associated with cloverleaf skull. Mutations affecting different functional domains of FGFR3 cause different forms of this lethal disorder.
Defects in FGFR3 are a cause of hypochondroplasia (HCH) [MIM:146000]. HCH is an autosomal dominant disease and is characterized by disproportionate short stature. It resembles achondroplasia, but with a less severe phenotype.
Defects in FGFR3 are a cause of susceptibility to bladder cancer (BLC) [MIM:109800]. A malignancy originating in tissues of the urinary bladder. It often presents with multiple tumors appearing at different times and at different sites in the bladder. Most bladder cancers are transitional cell carcinomas. They begin in cells that normally make up the inner lining of the bladder. Other types of bladder cancer include squamous cell carcinoma (cancer that begins in thin, flat cells) and adenocarcinoma (cancer that begins in cells that make and release mucus and other fluids). Bladder cancer is a complex disorder with both genetic and environmental influences. Note=Somatic mutations can constitutively activate FGFR3.
Defects in FGFR3 are a cause of cervical cancer (CERCA) [MIM:603956]. A malignant neoplasm of the cervix, typically originating from a dysplastic or premalignant lesion previously present at the active squamocolumnar junction. The transformation from mild dysplastic to invasive carcinoma generally occurs slowly within several years, although the rate of this process varies widely. Carcinoma in situ is particularly known to precede invasive cervical cancer in most cases. Cervical cancer is strongly associated with infection by oncogenic types of human papillomavirus.
Defects in FGFR3 are the cause of camptodactyly tall stature and hearing loss syndrome (CATSHL syndrome) [MIM:610474]. CATSHL syndrome is an autosomal dominant syndrome characterized by permanent and irreducible flexion of one or more fingers of the hand and/or feet, tall stature, scoliosis and/or a pectus excavatum, and hearing loss. Affected individuals have developmental delay and/or mental retardation, and several of these have microcephaly. Radiographic findings included tall vertebral bodies with irregular borders and broad femoral metaphyses with long tubular shafts. On audiological exam, each tested member have bilateral sensorineural hearing loss and absent otoacoustic emissions. The hearing loss was congenital or developed in early infancy, progressed variably in early childhood, and range from mild to severe. Computed tomography and magnetic resonance imaging reveal that the brain, middle ear, and inner ear are structurally normal.
Defects in FGFR3 are a cause of multiple myeloma (MM) [MIM:254500]. MM is a malignant tumor of plasma cells usually arising in the bone marrow and characterized by diffuse involvement of the skeletal system, hyperglobulinemia, Bence-Jones proteinuria and anemia. Complications of multiple myeloma are bone pain, hypercalcemia, renal failure and spinal cord compression. The aberrant antibodies that are produced lead to impaired humoral immunity and patients have a high prevalence of infection. Amyloidosis may develop in some patients. Multiple myeloma is part of a spectrum of diseases ranging from monoclonal gammopathy of unknown significance (MGUS) to plasma cell leukemia. Note=A chromosomal aberration involving FGFR3 is found in

multiple myeloma. Translocation t(4;14)(p16.3;q32.3) with the IgH locus.
Defects in FGFR3 are a cause of lacrimo-auriculo-dento-digital syndrome (LADDS) [MIM:149730]; also known as Levy-Hollister syndrome. LADDS is a form of ectodermal dysplasia, a heterogeneous group of disorders due to abnormal development of two or more ectodermal structures. LADDS is an autosomal dominant syndrome characterized by aplastic/hypoplastic lacrimal and salivary glands and ducts, cup-shaped ears, hearing loss, hypodontia and enamel hypoplasia, and distal limb segments anomalies. In addition to these cardinal features, facial dysmorphism, malformations of the kidney and respiratory system and abnormal genitalia have been reported. Craniosynostosis and severe syndactyly are not observed.
Defects in FGFR3 are a cause of keratinocytic non-epidermolytic nevus (KNEN) [MIM:162900]; also known as pigmented moles. Epidermal nevi of the common, non-organoid and non-epidermolytic type are benign skin lesions and may vary in their extent from a single (usually linear) lesion to widespread and systematized involvement. They may be present at birth or develop early during childhood.
Defects in FGFR3 are a cause of Muenke syndrome (MNKS) [MIM:602849]; also known as Muenke non-syndromic coronal craniosynostosis. MNKS is a condition characterized by premature closure of coronal suture of skull during development (coronal craniosynostosis), which affects the shape of the head and face. It may be uni- or bilateral. When bilateral, it is characterized by a skull with a small antero-posterior diameter (brachycephaly), often with a decrease in the depth of the orbits and hypoplasia of the maxillae. Unilateral closure of the coronal sutures leads to flattening of the orbit on the involved side (plagiocephaly). The intellect is normal. In addition to coronal craniosynostosis some affected individuals show skeletal abnormalities of hands and feet, sensorineural hearing loss, mental retardation and respiratory insufficiency.
Defects in FGFR3 are a cause of keratosis seborrheic (KERSEB) [MIM:182000]. A common benign skin tumor. Seborrheic keratoses usually begin with the appearance of one or more sharply defined, light brown, flat macules. The lesions may be sparse or numerous. As they initially grow, they develop a velvety to finely verrucous surface, followed by an uneven warty surface with multiple plugged follicles and a dull or lackluster appearance.

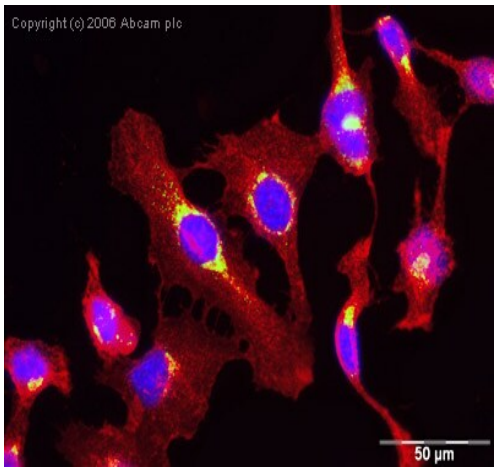
Sequence similarities

Belongs to the protein kinase superfamily. Tyr protein kinase family. Fibroblast growth factor receptor subfamily.
Contains 3 Ig-like C2-type (immunoglobulin-like) domains.
Contains 1 protein kinase domain.

Cellular localization

Membrane.

Images



Immunocytochemistry/ Immunofluorescence - Anti-FGFR3 antibody (ab10649)

ICC/IF image of ab10649 stained human HeLa cells. The cells were methanol fixed (5 min) and incubated with the antibody (ab10649, 1µg/ml) for 1h at room temperature. The secondary antibody (green) was Alexa Fluor® 488 goat anti-rabbit IgG (H+L) used at a 1/1000 dilution for 1h. Image-iT^{TM} FX Signal Enhancer was used as the primary blocking agent, 5% BSA (in TBS-T) was used for all other blocking steps. DAPI was used to stain the cell nuclei (blue). Alexa Fluor® 594 WGA was used to label plasma membranes (red).

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