SunLong Biotech Co.,LTD
Tel: 0086-571-56623320 Fax:0086-571-56623318
E-mail:sales@sunlongbiotech.com
www.sunlongbiotech.com

Rabbit Anti-FGFR2/AF350 Conjugated antibody

SL0675R-AF350

Product Name	Anti-FGFR2/AF350
Chinese Name	AF350 标记的成纤维细胞生长因子受体 2 抗体
Alias	KGFR; KSAM; Bacteria expressed kinase; BEK; BEK fibroblast growth factor receptor; BFR 1; BFR1; CD 332; CD332; CD332 antigen; CEK 3; CEK3; CFD 1; CFD1; Craniofacial dysostosis 1; Crouzon syndrome; ECT 1; ECT 1; ECT1; FGF receptor; FGFR 2; FGFR-2; Fgfr2; FGFR2_HUMAN; Fibroblast growth factor receptor 2; Hydroxyaryl protein kinase; Hydroxyaryl protein kinase; Jackson Weiss syndrome; JWS; JWS antibody K SAM; K sam protein; K sam protein; K-sam ; Keratinocyte growth factor receptor 2; Keratinocyte growth factor receptor; Pfeiffer syndrome; Protein tyrosine kinase receptor like 14; TK14; TK25; Tyrosylprotein kinase; Tyrosylprotein kinase.
Research Area	Tumour Cardiovascular immunology Neurobiology Signal transduction Stem cells Growth factors and hormones Kinases and Phosphatases
Immunogen Species	Rabbit
Clonality	Polyclonal
React Species	Human,Mouse(predicted:Rat)
Applications	Flow-Cyt=1μg/Test,IF=1:100-500,ICC/IF=1:100-500 not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight	89kDa
Form	Lyophilized or Liquid
Concentration	1mg/ml
immunogen	KLH conjugated synthetic peptide derived from human FGFR2
Lsotype	IgG
Purification	affinity purified by Protein A
Storage Buffer	1M TBS(pH7.4) with 1% BSA, 3% Proclin300 and 50% Glycerol
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and

for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 1M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Product Detail

background:
Fibroblast growth factors (FGFs) are members of a large family of structurally related polypeptides that are potent physiological regulators of growth and differentiation for a wide variety of cells of mesodermal, ectodermal and endodermal origin. Four genes encoding for high affinity cell surface FGF receptors (FGFRs) have been identified: FGFR1, FGFR2, FGFR3 and FGFR4. FGFRs are members of the tyrosine kinase family of growth factor receptors. FGFR2 is highly expressed in developing human tissues including the brain, choroids plexus, lung etc. Alternative names: Bacteria expressed kinase; BEK; BFR 1; BFR1; CD 332; CD332; CD332 antigen; CEK 3; CEK3; CFD 1; CFD1; Craniofacial dysostosis 1; Crouzon syndrome; ECT 1; ECT1; FGFR 2; Fibroblast growth factor receptor 2; Hydroxyaryl protein kinase; Jackson Weiss syndrome; JWS; K SAM; K sam protein; Keratinocyte growth factor receptor 2; Keratinocyte growth factor receptor; KGFR; KSAM; Pfeiffer syndrome; Protein tyrosine kinase receptor like 14; TK14; TK25; Tyrosylprotein kinase.

Function:
Tyrosine-protein kinase that acts as cell-surface receptor for fibroblast growth factors and plays an essential role in the regulation of cell proliferation, differentiation, migration and apoptosis, and in the regulation of embryonic development. Required for normal embryonic patterning, trophoblast function, limb bud development, lung morphogenesis, osteogenesis and skin development. Plays an essential role in the regulation of osteoblast differentiation, proliferation and apoptosis, and is required for normal skeleton development. Promotes cell proliferation in keratinocytes and immature osteoblasts, but promotes apoptosis in differentiated osteoblasts. Phosphorylates PLCG1, FRS2 and PAK4. Ligand binding leads to the activation of several signaling cascades. Activation of PLCG1 leads to the production of the cellular signaling molecules diacylglycerol and inositol 1,4,5-trisphosphate. Phosphorylation of FRS2 triggers recruitment of GRB2, GAB1, PIK3R1 and SOS1, and mediates activation of RAS, MAPK1/ERK2, MAPK3/ERK1 and the MAP kinase signaling pathway, as well as of the AKT1 signaling pathway. FGFR2 signaling is down-regulated by ubiquitination, internalization and degradation. Mutations that lead to constitutive kinase activation or impair normal FGFR2 maturation, internalization and degradation lead to aberrant signaling. Over-expressed FGFR2 promotes activation of STAT1.

Subunit:
Monomer. Homodimer after ligand binding. Interacts predominantly with

FGF1 and FGF2, but can also interact with FGF3, FGF4, FGF6, FGF7, FGF8, FGF9, FGF10, FGF17, FGF18 and FGF22 (in vitro). Ligand specificity is determined by tissue-specific expression of isoforms, and differences in the third Ig-like domain are crucial for ligand specificity. Isoform 1 has high affinity for FGF1 and FGF2, but low affinity for FGF7. Isoform 3 has high affinity for FGF1 and FGF7, and has much higher affinity for FGF7 than isoform 1 (in vitro). Affinity for fibroblast growth factors (FGFs) is increased by heparan sulfate glycosaminoglycans that function as coreceptors. Likewise, KLB increases the affinity for FGF19 and FGF21. Interacts with PLCG1, GRB2 and PAK4.

Subcellular Location:
Cell membrane; Single-pass type I membrane protein. Golgi apparatus. Cytoplasmic vesicle. Note=Detected on osteoblast plasma membrane lipid rafts. After ligand binding, the activated receptor is rapidly internalized and degraded.
Isoform 1: Cell membrane; Single-pass type I membrane protein. Note=After ligand binding, the activated receptor is rapidly internalized and degraded.
Isoform 3: Cell membrane; Single-pass type I membrane protein. Note=After ligand binding, the activated receptor is rapidly internalized and degraded.

Post-translational modifications:
N-glycosylated in the endoplasmic reticulum. The N-glycan chains undergo further maturation to an Endo H-resistant form in the Golgi apparatus. Ubiquitinated. FGFR2 is rapidly ubiquitinated after autophosphorylation, leading to internalization and degradation. Subject to degradation both in lysosomes and by the proteasome.

DISEASE:
Defects in FGFR2 are the cause of Crouzon syndrome (CS) [MIM:123500]; also called craniofacial dysostosis type I (CFD1). CS is an autosomal dominant syndrome characterized by craniosynostosis (premature fusion of the skull sutures), hypertelorism, exophthalmos and external strabismus, parrot-beaked nose, short upper lip, hypoplastic maxilla, and a relative mandibular prognathism.
Defects in FGFR2 are a cause of Jackson-Weiss syndrome (JWS) [MIM:123150]. JWS is an autosomal dominant craniosynostosis syndrome characterized by craniofacial abnormalities and abnormality of the feet: broad great toes with medial deviation and tarsal-metatarsal coalescence. Defects in FGFR2 are a cause of Apert syndrome (APRS) [MIM:101200]; also known as acrocephalosyndactyly type 1 (ACS1). APRS is a syndrome characterized by facio-cranio-synostosis, osseous and membranous syndactyly of the four extremities, and midface hypoplasia. The craniosynostosis is bicoronal and results in acrocephaly of brachysphenocephalic type. Syndactyly of the fingers

and toes may be total (mitten hands and sock feet) or partial affecting the second, third, and fourth digits. Intellectual deficit is frequent and often severe, usually being associated with cerebral malformations.
Defects in FGFR2 are a cause of Pfeiffer syndrome (PS) [MIM:101600]; also known as acrocephalosyndactyly type V (ACS5). PS is characterized by craniosynostosis (premature fusion of the skull sutures) with deviation and enlargement of the thumbs and great toes, brachymesophalangy, with phalangeal ankylosis and a varying degree of soft tissue syndactyly. Three subtypes of Pfeiffer syndrome have been described: mild autosomal dominant form (type 1); cloverleaf skull, elbow ankylosis, early death, sporadic (type 2); craniosynostosis, early demise, sporadic (type 3).
Defects in FGFR2 are the cause of Beare-Stevenson cutis gyrata syndrome (BSCGS) [MIM:123790]. BSCGS is an autosomal dominant condition is characterized by the furrowed skin disorder of cutis gyrata, acanthosis nigricans, craniosynostosis, craniofacial dysmorphism, digital anomalies, umbilical and anogenital abnormalities and early death.
Defects in FGFR2 are the cause of familial scaphocephaly syndrome (FSPC) [MIM:609579]; also known as scaphocephaly with maxillary retrusion and mental retardation. FSPC is an autosomal dominant craniosynostosis syndrome characterized by scaphocephaly, macrocephaly, hypertelorism, maxillary retrusion, and mild intellectual disability. Scaphocephaly is the most common of the craniosynostosis conditions and is characterized by a long, narrow head. It is due to premature fusion of the sagittal suture or from external deformation.
Defects in FGFR2 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 FGFR2 are the cause of Antley-Bixler syndrome (ABS) [MIM:207410]. ABS is a multiple congenital anomaly syndrome characterized by craniosynostosis, radiohumeral synostosis, midface hypoplasia, malformed ears, arachnodactyly and multiple joint contractures. ABS is a heterogeneous disorder and occurs with and without abnormal genitalia in both sexes.

Similarity:
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.

Database links:

Entrez Gene: 2263 Human

Entrez Gene: 14183 Mouse

Entrez Gene: 25022 Rat

Omim: 176943 Human

SwissProt: P21802 Human

SwissProt: P21803 Mouse

Unigene: 533683 Human

Unigene: 16340 Mouse

Unigene: 12732 Rat

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

KGFR 又称 FGFR2（Fibroblast Growth Factor Receptor 2）成纤维细胞生长因子受体又称：纤维母细胞生长因子受体 2，是 FGFRs 家族的一员。不同的 FGFR 对 FGF 亲和力不同，在组织的分布也不一样。FGFR-2 对细胞的增殖、分化、血管生成、胚胎及骨骼发育和在与生长发育相关的进程中起着十分重要的作用.