

Anti-FGFR3 antibody [EPR2304(3)] ab133644

KO 評価済 リコンビナント RabMAb

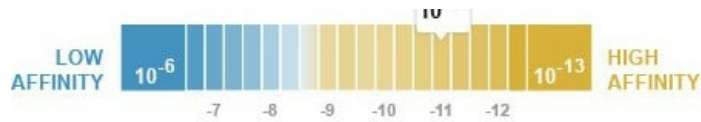
★★★★★ [2 Abreviews](#) [12 References](#) [画像数 4](#)

製品の概要

製品名	Anti-FGFR3 antibody [EPR2304(3)]
製品の詳細	Rabbit monoclonal [EPR2304(3)] to FGFR3
由来種	Rabbit
特異性	We have tested this antibody in positive Mouse and Rat tissue lysates in WB and could not obtain a specific band for FGFR3. Please get in touch with our Scientific Support team if you wish to share any data regarding this antibody's cross-reactivity in Mouse and Rat species.
アプリケーション	適用あり: WB 適用なし: Flow Cyt, ICC/IF or IHC-P
種交差性	交差種: Human
免疫原	Synthetic peptide within Human FGFR3 aa 1-100 (extracellular). The exact sequence is proprietary. Database link: P22607 (Peptide available as ab195871)
ポジティブ・コントロール	WB: K562, HepG2 and HEK293 lysates.
特記事項	This product is a recombinant monoclonal antibody, which offers several advantages including: <ul style="list-style-type: none">- 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 .

製品の特性

製品の状態	Liquid
保存方法	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C. Stable for 12 months at -20°C.
解離定数 (K _D 値)	K _D = 1.62 x 10 ⁻¹¹ M



[Learn more about \$K_D\$](#)

バッファー	pH: 7.20 Preservative: 0.01% Sodium azide Constituents: 59% PBS, 0.05% BSA, 40% Glycerol
精製度	Protein A purified
ポリモノ	モノクローナル
クローン名	EPR2304(3)
アイソタイプ	IgG

アプリケーション

The Abpromise guarantee **Abpromise保証は、次のテスト済みアプリケーションにおけるab133644の使用に適用されます**
 アプリケーションノートには、推奨の開始希釈率がありますが、適切な希釈率につきましてはご検討ください。

アプリケーション	Abreviews	特記事項
WB	★★★★★ (1)	1/1000 - 1/2000. Predicted molecular weight: 88 kDa. Abcam recommends to block in 3% non-fat milk.

追加情報 Is unsuitable for Flow Cyt, ICC/IF or IHC-P.

ターゲット情報

機能	Receptor for acidic and basic fibroblast growth factors. Preferentially binds FGF1.
組織特異性	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.
関連疾患	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.

配列類似性

Belongs to the protein kinase superfamily. Tyr protein kinase family. Fibroblast growth factor receptor subfamily.

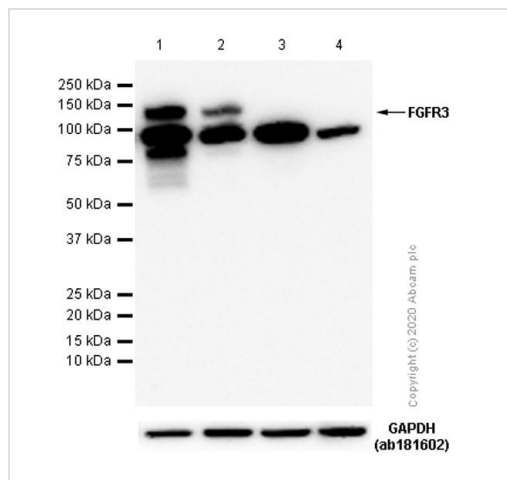
Contains 3 Ig-like C2-type (immunoglobulin-like) domains.

Contains 1 protein kinase domain.

細胞内局在

Membrane.

画像



Western blot - Anti-FGFR3 antibody [EPR2304(3)] (ab133644)

All lanes : Anti-FGFR3 antibody [EPR2304(3)] (ab133644) at 1/2000 dilution

Lane 1 : HepG2 (Human hepatocellular carcinoma epithelial cell) whole cell lysates

Lane 2 : K-562 (Human chronic myelogenous leukemia lymphoblast) whole cell lysates

Lane 3 : MCF7 (Human breast adenocarcinoma epithelial cell) whole cell lysates

Lane 4 : IM-9 (Human multiple myeloma B Lymphoblast) whole cell lysates

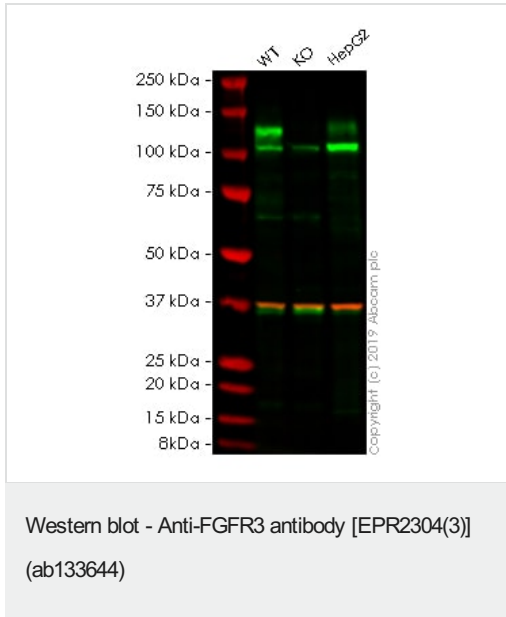
Lysates/proteins at 20 µg per lane.

Secondary

All lanes : Goat Anti-Rabbit IgG, (H+L), Peroxidase conjugated (ab97051) at 1/20000 dilution

Predicted band size: 88 kDa

Blocking/Diluting buffer and concentration: 5% NFDM/TBST



All lanes : Anti-FGFR3 antibody [EPR2304(3)] (ab133644) at 1/1000 dilution

Lane 1 : Wild-type HEK-293 (Human epithelial cell line from embryonic kidney) whole cell lysate

Lane 2 : FGFR3 knockout HEK-293 (Human epithelial cell line from embryonic kidney) whole cell lysate

Lane 3 : Hep G2 (Human liver hepatocellular carcinoma cell line) whole cell lysate

Lysates/proteins at 20 µg per lane.

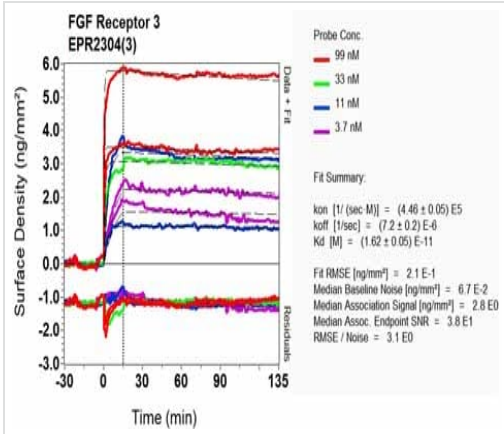
Performed under reducing conditions.

Predicted band size: 88 kDa

Observed band size: 125 kDa

Lanes 1 - 3: Merged signal (red and green). Green - ab133644 observed at 125kDa. Red - loading control, **ab9484**, observed at 37 kDa.

ab133644 was shown to recognize in wild-type HEK-293 cells as signal was lost at the expected MW in FGFR3 knockout cells. Additional cross-reactive bands were observed in the wild-type and knockout cells. Wild-type and FGFR3 knockout samples were subjected to SDS-PAGE. Ab133644 and **ab9484** (Mouse anti-GAPDH loading control) were incubated overnight at 4°C at 1/1000 dilution and 1/20000 dilution respectively. Blots were developed with Goat anti-Rabbit IgG H&L (IRDye® 800CW) preabsorbed **ab216773** and Goat anti-Mouse IgG H&L (IRDye® 680RD) preabsorbed **ab216776** secondary antibodies at 1/20000 dilution for 1 hour at room temperature before imaging.



Equilibrium dissociation constant (K_D)

Learn more about K_D

[Click here to learn more about \$K_D\$](#)

OI-RD Scanning - Anti-FGFR3 antibody
 [EPR2304(3)] (ab133644)

Why choose a recombinant antibody?

Research with confidence
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 Recombinant technology

Success from the first experiment
 Confirmed specificity

Ethical standards compliant
 Animal-free production

Anti-FGFR3 antibody [EPR2304(3)] (ab133644)

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