SANTA CRUZ BIOTECHNOLOGY, INC.

ALK-1 (E-11): sc-518147

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

Hereditary hemorrhagic telangiectasia (HHT) is an autosomal dominant disorder characterized by vascular abnormalities such as dilated vessels, hemorrhages, liver and lung congestion, and brain or heart ischemia. Mutations in two genes, Endoglin (also designated CD105) and ALK-1 (activin receptor-like kinase 1, also designated TGFβ superfamily RI), are responsible for HHT. Endoglin is mutated in HHT1, and ALK-1 is mutated in HHT2, both of which are thought to be caused by haploinsufficiency. Endoglin and ALK-1 are type III and type I members of the TGFβ receptor superfamily, respectively, that are expressed on vascular endothelial cells. Endoglin can only bind ligands of the TGFβ superfamily via association with the respective ligand binding receptors for TGFβ1, TGFβ3, activin-A, BMP-2 and BMP-7. ALK-1 preferentially binds TGFβ1 and is expressed in bone marrow stromal cells, lung, brain, kidney and spleen.

REFERENCES

1. Wu, X., et al. 1995. Cloning and characterization of the murine activin receptor like kinase-1 (ALK-1) homolog. Biochem. Biophys. Res. Commun. 216: 78-83.
2. Altomonte, M., et al. 1996. Expression and structural features of Endoglin (CD105), a transforming growth factor β1 and β3 binding protein, in human melanoma. Br. J. Cancer 74: 1586-1591.
3. Gallione, C.J., et al. 1998. Mutation and expression analysis of the Endoglin gene in hereditary hemorrhagic telangiectasia reveals null alleles. Hum. Mutat. 11: 286-294.
4. Klaus, D.J., et al. 1998. Novel missense and frameshift mutations in the activin receptor-like kinase-1 gene in hereditary hemorrhagic telangiectasia. Hum. Mutat. 12: 137.
5. Bourdeau, A., et al. 2000. Endoglin-deficient mice, a unique model to study hereditary hemorrhagic telangiectasia. Trends Cardiovasc. Med. 10: 279-285.
6. Azuma, H. 2000. Genetic and molecular pathogenesis of hereditary hemorrhagic telangiectasia. J. Med. Invest. 47: 81-90.
7. Bourdeau, A., et al. 2001. Potential role of modifier genes influencing transforming growth factor-β1 levels in the development of vascular defects in Endoglin heterozygous mice with hereditary hemorrhagic telangiectasia. Am. J. Pathol. 158: 2011-2020.

CHROMOSOMAL LOCATION

Genetic locus: ACVRL1 (human) mapping to 12q13.13.

SOURCE

ALK-1 (E-11) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 55-73 of ALK-1 of human origin.

PRODUCT

Each vial contains 200 µg IgM kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

ALK-1 (E-11) is recommended for detection of ALK-1 of human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for ALK-1 siRNA (h): sc-40212, ALK-1 shRNA Plasmid (h): sc-40212-SH and ALK-1 shRNA (h) Lentiviral Particles: sc-40212-V.

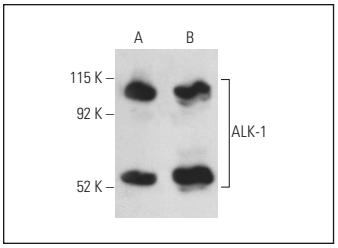
Molecular Weight of ALK-1: 53 kDa.

Positive Controls: human brain extract: sc-364375 or human kidney extract: sc-363764.

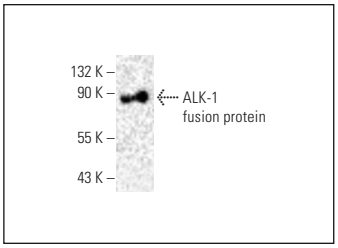
RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein L-Agarose: sc-2336 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

DATA



ALK-1 (E-11): sc-518147. Western blot analysis of ALK-1 expression in human brain (**A**) and human kidney (**B**) tissue extracts. Detection reagent used: m-IgGκ BP-HRP: sc-516102.



ALK-1 (E-11): sc-518147. Western blot analysis of human recombinant ALK-1 fusion protein. Detection reagent used: m-IgGκ BP-HRP: sc-516102.

STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

RESEARCH USE

For research use only, not for use in diagnostic procedures.

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.