

ALK-1 (C-20): sc-19547

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 TGF β 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 TGF β receptor superfamily, respectively, that are expressed on vascular endothelial cells. Endoglin can only bind ligands of the TGF β superfamily via association with the respective ligand binding receptors for TGF β 1, TGF β 3, Activin-A, BMP-2 and BMP-7. ALK-1 preferentially binds TGF β 1 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 β 1 and β 3 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.

CHROMOSOMAL LOCATION

Genetic locus: ACVRL1 (human) mapping to 12q13.13; Acvrl1 (mouse) mapping to 15 F2.

SOURCE

ALK-1 (C-20) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus of ALK-1 of human origin.

PRODUCT

Each vial contains 200 μ g IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-19547 P, (100 μ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

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.

APPLICATIONS

ALK-1 (C-20) is recommended for detection of ALK-1 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500), immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

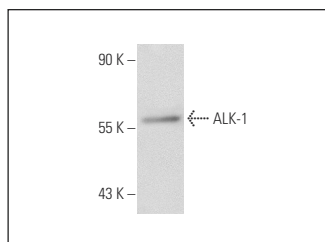
ALK-1 (C-20) is also recommended for detection of ALK-1 in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for ALK-1 siRNA (h): sc-40212, ALK-1 siRNA (m): sc-40213, ALK-1 shRNA Plasmid (h): sc-40212-SH, ALK-1 shRNA Plasmid (m): sc-40213-SH, ALK-1 shRNA (h) Lentiviral Particles: sc-40212-V and ALK-1 shRNA (m) Lentiviral Particles: sc-40213-V.

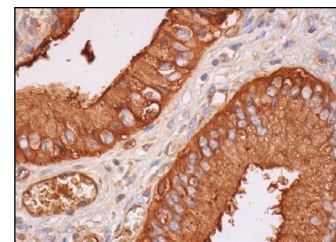
Molecular Weight of ALK-1: 53 kDa.

Positive Controls: human platelet extract: sc-363773.

DATA



ALK-1 (C-20): sc-19547. Western blot analysis of ALK-1 expression in human platelet extract.



ALK-1 (C-20): sc-19547. Immunoperoxidase staining of formalin fixed, paraffin-embedded human gall bladder tissue showing cytoplasmic and membrane staining of glandular cells.

SELECT PRODUCT CITATIONS

1. Medici, D., et al. 2010. Conversion of vascular endothelial cells into multi-potent stem-like cells. *Nat. Med.* 16: 1400-1406.

PROTOCOLS

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

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Try **ALK-1 (RM0015-1B03): sc-101556**, our highly recommended monoclonal alternative to ALK-1 (C-20).