

## ALK-1 (D-20): sc-19546

### 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 $\beta$  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 $\beta$  receptor superfamily, respectively, that are expressed on vascular endothelial cells. Endoglin can only bind ligands of the TGF $\beta$  superfamily via association with the respective ligand binding receptors for TGF $\beta$ 1, TGF $\beta$ 3, Activin-A, BMP-2 and BMP-7. The human ALK-1 gene encodes two protein species which exist as a result of either glycosylation or alternative splicing events. ALK-1 preferentially binds TGF $\beta$ 1 and is expressed in bone marrow stromal cells, lung, brain, kidney and spleen.

### CHROMOSOMAL LOCATION

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

### SOURCE

ALK-1 (D-20) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of ALK-1 of mouse origin.

### PRODUCT

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

Blocking peptide available for competition studies, sc-19546 P, (100  $\mu$ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

### RESEARCH USE

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

### APPLICATIONS

ALK-1 (D-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), 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).

ALK-1 (D-20) is also recommended for detection of ALK-1 in additional species, including equine.

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.

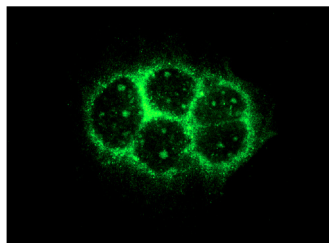
Molecular Weight of ALK-1: 53 kDa.

Positive Controls: human platelet extract: sc-363773.

### STORAGE

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

### DATA



ALK-1 (D-20): sc-19546. Immunofluorescence staining of methanol-fixed JAR cells showing membrane localization.

### SELECT PRODUCT CITATIONS

1. König, H.G., et al. 2005. TGF $\beta$ 1 activates two distinct type I receptors in neurons: implications for neuronal NF $\kappa$ B signaling. *J. Cell Biol.* 168: 1077-1086.
2. Castaneres, C., et al. 2007. Signaling by ALK5 mediates TGF- $\beta$ -induced ET-1 expression in endothelial cells: a role for migration and proliferation. *J. Cell Sci.* 120: 1256-1266.
3. Yao, Y., et al. 2007. Activin-like kinase receptor 1 (ALK1) in atherosclerotic lesions and vascular mesenchymal cells. *Cardiovasc. Res.* 74: 279-289.
4. Braun, A., et al. 2007. Paucity of pericytes in germinal matrix vasculature of premature infants. *J. Neurosci.* 27: 12012-12024.
5. Velasco, S., et al. 2008. L- and S-endoglin differentially modulate TGF $\beta$ 1 signaling mediated by ALK-1 and ALK-5 in L6E9 myoblasts. *J. Cell Sci.* 121: 913-919.
6. Wang, S., et al. 2009. Diabetes-relevant regulation of cultured blood outgrowth endothelial cells. *Microvasc. Res.* 78: 174-179.
7. Blaney Davidson, E.N., et al. 2009. Increase in ALK1/ALK5 ratio as a cause for elevated MMP-13 expression in osteoarthritis in humans and mice. *J. Immunol.* 182: 7937-7945.

### PROTOCOLS

See our web site at [www.scbt.com](http://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 (D-20).