# SANTA CRUZ BIOTECHNOLOGY, INC.

# ALK-1 (RM0015-1B03): sc-101556



#### The Fower to quest

# 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 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  $\beta$ 1 and  $\beta$ 3 binding protein, in human melanoma. Br. J. Cancer 74: 1586-1591.
- 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.
- 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.
- 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- $\beta$ 1 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; Acvrl1 (mouse) mapping to 15 F2.

# SOURCE

ALK-1 (RM0015-1B03) is a rat monoclonal antibody raised against the extracellular domain of ALK-1 of mouse origin.

# PRODUCT

Each vial contains 100  $\mu g~lg G_2$  in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

# **APPLICATIONS**

ALK-1 (RM0015-1B03) 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 µg per 100-500 µg of total protein (1 ml of cell lysate)] and immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500).

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 (RM0015-1B03): sc-101556. Western blot analysis of ALK-1 expression in human platelet extract.

#### SELECT PRODUCT CITATIONS

- Zhu, Q., et al. 2013. SnoN facilitates ALK1-Smad1/5 signaling during embryonic angiogenesis. J. Cell Biol. 202: 937-950.
- Qu, Z., et al. 2020. High-dose TGF-β1 degrades human nucleus pulposus cells via ALK1-Smad1/5/8 activation. Exp. Ther. Med. 20: 3661-3668.
- Yuan, Y., et al. 2021. A pulmonary vascular model from endothelialized whole organ scaffolds. Front. Bioeng. Biotechnol. 9: 760309.
- Lösch, L., et al. 2023. GDF15 promotes the osteogenic cell fate of periodontal ligament fibroblasts, thus affecting their mechanobiological response. Int. J. Mol. Sci. 24: 10011.

#### **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.