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. 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 TGFβ1 and is expressed in bone marrow stromal cells, lung, brain, kidney and spleen.

CHROMOSOMAL LOCATION

Genetic locus: ACVRL1 (human) mapping to 12q13.13; Acrv1 (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 µg 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 µg 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

SELECT PRODUCT CITATIONS


PROTOCOLS

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

MONOS Satisfaction Guaranteed

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