SANTA CRUZ BIOTECHNOLOGY, INC.

Endoglin (8E11): sc-53999

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.

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. Mutations in brief no. 164. Online. 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. Gallione, C.J., et al. 2000. Two common Endoglin mutations in families with hereditary hemorrhagic telangiectasia in the Netherlands Antilles: evidence for a founder effect. Hum. Genet. 107: 40-44.

CHROMOSOMAL LOCATION

Genetic locus: ENG (human) mapping to 9q34.11.

SOURCE

Endoglin (8E11) is a mouse monoclonal antibody raised against monocytic U937 phorbol ester-treated cells of human origin.

STORAGE

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

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.

Endoglin (8E11) is available conjugated to either phycoerythrin (sc-53999 PE) or fluorescein (sc-53999 FITC), 200 µg/ml, for IF, IHC(P) and FCM.

APPLICATIONS

Endoglin (8E11) is recommended for detection of Endoglin of 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 flow cytometry (1 µg per 1 x 10^6 cells).

Suitable for use as control antibody for Endoglin siRNA (h): sc-35302, Endoglin shRNA Plasmid (h): sc-35302-SH and Endoglin shRNA (h) Lentiviral Particles: sc-35302-V.

Molecular Weight of reduced Endoglin: 84 kDa.

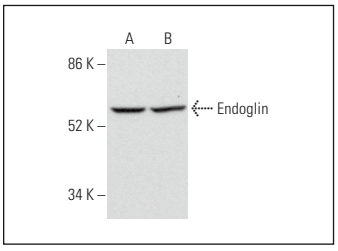
Molecular Weight of non-reduced Endoglin: 130 kDa.

Positive Controls: A549 cell lysate: sc-2413, ECV304 cell lysate: sc-2269 or HL-60 whole cell lysate: sc-2209.

RECOMMENDED SUPPORT 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).

DATA



Endoglin (8E11): sc-53999. Western blot analysis of Endoglin expression in ECV304 (**A**) and A549 (**B**) whole cell lysates.

RESEARCH USE

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