**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: ENG (human) mapping to 9q34.11; Eng (mouse) mapping to 2 B.

**SOURCE**

Endoglin (P4A4) is a mouse monoclonal antibody raised against Endoglin of human origin.

**PRODUCT**

Each vial contains 200 µg IgG₂κ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin. Also available azide-free for activation of T cell binding to endothelium, sc-20072 L, 200 µg/0.1 ml.

Endoglin (P4A4) is available conjugated to agarose (sc-20072 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-20072 HRP), 200 µg/ml, for WB, (HCPP) and ELISA; to either phycoerythrin (sc-20072 PE), fluorescein (sc-20072 FITC), Alexa Fluor® 488 (sc-20072 AF488), Alexa Fluor® 546 (sc-20072 AF546), Alexa Fluor® 594 (sc-20072 AF594) or Alexa Fluor® 647 (sc-20072 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-20072 AF680) or Alexa Fluor® 790 (sc-20072 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

**APPLICATIONS**

Endoglin (P4A4) is recommended for detection of Endoglin dimer under non-reducing conditions, and Endoglin monomer under reducing conditions 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).


Molecular Weight of reduced Endoglin: 84 kDa.

Molecular Weight of non-reduced Endoglin: 130 kDa.

**STORAGE**

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

**DATA**

Endoglin (P4A4): sc-20072. Western blot analysis of Endoglin expression in human spleen (A) and human kidney (B) tissue extracts under non-reducing conditions.

**SELECT PRODUCT CITATIONS**


**RESEARCH USE**

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

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