bs-7610R

[Primary Antibody]

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ETHE1 Rabbit pAb

- DATASHEET -

Host: Rabbit Isotype: IgG

Clonality: Polyclonal

GenelD: 23474 **SWISS:** 095571

Target: ETHE1

Immunogen: KLH conjugated synthetic peptide derived from human ETHE1:

191-254/254.

Purification: affinity purified by Protein A

Concentration: 1mg/ml

Storage: 0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50%

Glycerol.

Shipped at 4°C. Store at -20°C for one year. Avoid repeated

freeze/thaw cycles.

Background: Probably plays an important role in metabolic homeostasis in

mitochondria. May function as a nuclear-cytoplasmic shuttling protein that binds transcription factor RELA/NFKB3 in the nucleus and exports it to the cytoplasm. Suppresses p53-induced apoptosis

by preventing nuclear localization of RELA.

Involvement in disease:Defects in ETHE1 are a cause of ethylmalonic encephalopathy (EE) . EE is an autosomal recessive disorder characterized by neurodevelopmental delay and regression, recurrent petechiae, acrocyanosis, diarrhea, leading to death in the first decade of life. It is also associated with persistent

lactic acidemia and ethylmalonic and methylsuccinic aciduria.

Applications: ELISA (1:5000-10000)

Reactivity: (predicted: Human, Mouse,

Rabbit, Pig, Sheep, Cow,

Dog)

Predicted MW.: 26 kDa

Subcellular Cytoplasm , Nucleus