bs-7610R

[Primary Antibody]

ETHE1 Rabbit pAb



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- DATASHEET 400-901-9800		
Host: Rabbit	lsotype: IgG	Applications: ELISA (1:5000-10000)
Clonality: Polyclonal		Reactivity: (predicted: Human, Mouse,
GenelD: 23474	SWISS: 095571	Rabbit, Pig, Sheep, Cow,
Target: ETHE1		Dog)
Immunogen: KLH conjugated synthetic peptide derived from human ETHE1: 191-254/254.		Predicted 26 kDa
Purification: affinity purified by Protein A		Subcollular
Concentration: 1mg/ml		Subcellular Location: Cytoplasm ,Nucleus
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.		eus otosis ive ng to tent