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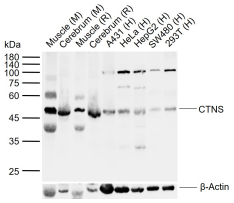
CTNS Rabbit pAb

Catalog Number:	bs-12932R
Target Protein:	CTNS
Concentration:	1mg/ml
Form:	Liquid
Host:	Rabbit
Clonality:	Polyclonal
Isotype:	IgG
Applications:	WB (1:500-2000)
Reactivity:	Human, Mouse, Rat (predicted:Rabbit)
Predicted MW:	42 kDa
Entrez Gene:	1497
Swiss Prot:	O60931
Source:	KLH conjugated synthetic peptide derived from human CTNS/Cystinosin: 231-330/367.
Purification:	affinity purified by Protein A
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:	Cystinosis is an autosomal recessive disorder resulting from defective lysosomal transport of cystine and present at birth as a failure to thrive, rickets and proximal renal tubular acidosis. The human CTNS gene on chromosome 17p13 encodes the protein Cystinosin, and mutations in CTNS are responsible for nephropathic cystinosis. The CTNS promoter contains an Sp1 binding element. Cystinosin is an integral membrane protein containing 7 transmembrane domains that functions as a H+-driven transporter responsible for cystine export from lysosomes. In humans, Cystinosin is expressed abundantly in pancreas, kidney (mature and fetal), and skeletal muscle. The mouse homolog to CTNS encodes a protein which is expressed in all tissues except skeletal muscle. In the cell, Cystinosin co-localizes with LAMP-2 to lysosomes. A C-terminal GYDQL sorting motif within Cystinosin is critical for lysosomal localization.

VALIDATION IMAGES



Sample: Lane 1: Mouse Muscle tissue lysates Lane 2: Mouse Cerebrum tissue lysates Lane 3: Rat Muscle tissue lysates Lane 4: Rat Cerebrum tissue lysates Lane 5: Human A431 cell lysates Lane 6: Human HeLa cell lysates Lane 7: Human HepG2 cell lysates Lane 8: Human SW480 cell lysates Lane 9: Human 293T cell lysates Primary: Anti-CTNS (bs-12932R) at 1/1000 dilution Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution Predicted band size: 42 kDa Observed band size: 48 kDa