

[Primary Antibody]

CTNS Rabbit pAb



BioSS
ANTIBODIES

www.bioss.com.cn

sales@bioass.com.cn

techsupport@bioss.com.cn

400-901-9800

— DATASHEET

Host: Rabbit

Isotype: IgG

Clonality: Polyclonal

GeneID: 1497

SWISS: 060931

Target: CTNS

Immunogen: KLH conjugated synthetic peptide derived from human CTNS/Cystinosin: 231-330/367.

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: 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 Cystinosisin, and mutations in CTNS are responsible for nephropathic cystinosis. The CTNS promoter contains an Sp1 binding element. Cystinosisin is an integral membrane protein containing 7 transmembrane domains that functions as a H⁺-driven transporter responsible for cystine export from lysosomes. In humans, Cystinosisin 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, Cystinosisin co-localizes with LAMP-2 to lysosomes. A C-terminal GYDQL sorting motif within Cystinosisin is critical for lysosomal localization.

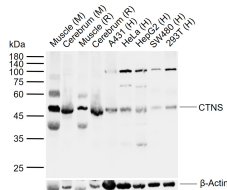
Applications: WB (1:500-2000)

Reactivity: Human, Mouse, Rat
(predicted: Rabbit)

Predicted MW.: 42 kDa

Subcellular Location: Cell membrane ,Cytoplasm

— 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