SANTA CRUZ BIOTECHNOLOGY, INC.

Cystinosin (H-100): sc-134899

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.

REFERENCES

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5. Cherqui, S., Kalatzis, V., Trugnan, G. and Antignac, C. 2001. The targeting of cystinosin to the lysosomal membrane requires a tyrosine-based signal and a novel sorting motif. J. Biol. Chem. 276: 13314-13321.

CHROMOSOMAL LOCATION

Genetic locus: CTNS (human) mapping to 17p13.2; Ctns (mouse) mapping to 11 B4.

SOURCE

Cystinosin (H-100) is a rabbit polyclonal antibody raised against amino acids 68-167 mapping within an internal region of Cystinosin of human origin.

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

STORAGE

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

APPLICATIONS

Cystinosin (H-100) is recommended for detection of Cystinosin 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)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Cystinosin (H-100) is also recommended for detection of Cystinosin in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for Cystinosin siRNA (h): sc-105264, Cystinosin siRNA (m): sc-142757, Cystinosin shRNA Plasmid (h): sc-105264-SH, Cystinosin shRNA Plasmid (m): sc-142757-SH, Cystinosin shRNA (h) Lentiviral Particles: sc-105264-V and Cystinosin shRNA (m) Lentiviral Particles: sc-142757-V.

Molecular Weight of Cystinosin: 42 kDa.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

RESEARCH USE

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

PROTOCOLS

See our web site at www.scbt.com or our catalog for detailed protocols and support products.