SANTA CRUZ BIOTECHNOLOGY, INC.

Cystinosin (F-12): sc-100703

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

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 17p13.3.

SOURCE

Cystinosin (F-12) is a mouse monoclonal antibody raised against recombinant Cystinosin of human origin.

PRODUCT

Each vial contains 100 µg IgG_{2a} kappa light chain 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

CCystinosin (F-12) is recommended for detection of Cystinosin of 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)] and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

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

Molecular Weight of Cystinosin: 42 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227.

RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml).

RESEARCH USE

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

PROTOCOLS

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