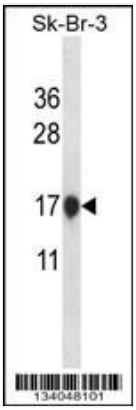
CSTB Antibody – Rabbit Polyclonal

PRODUCT OVERVIEW

Product Name	CSTB, Polyclonal Antibody
Full Product Name	CSTB, ID (CSTB, CST6, STFB, Cystatin-B, CPI-B, Liver thiol proteinase inhibitor, Stefin-B) (FITC)
Product Gene Name	anti-CSTB antibody
Product Synonym Names	CSTB; CST6; STFB; Cystatin-B; CPI-B; Liver thiol proteinase inhibitor; Stefin-B
Clonality	Polyclonal
Isotype	IgG
Host	Rabbit
Species Reactivity	Human
Form/Format	Supplied as a liquid in PBS, pH 7.2. No preservative added. Labeled with Fluorescein Isothiocyanate (FITC).
Purity/Purification	Purified by Protein A Affinity Chromatography.
Preparation and Storage	Store product at 4 degree C if to be used immediately within two weeks. For long-term storage, aliquot to avoid repeated freezing and thawing and store at -20 degree C. Aliquots are stable at -20 degree C for 12 months after receipt. Dilute required amount only prior to immediate use. Further dilutions can be made in assay buffer. Caution: FITC conjugates are sensitive to light. For maximum recovery of product, centrifuge the original vial after thawing and prior to removing the cap.
Molecular Weight	11,140 Da
Chromosome Location	Chromosome: 21; NC_000021.8 (45193546..45196256, complement). Location: 21q22.3
OMIM	254800
3D Structure	ModBase 3D Structure for P04080
Immunogen	CSTB antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 9-39 amino acids from the Central region of human CSTB.
Conjugate	FITC
Note	Preservative Free
Special Handling	Light sensitive
Product Categories/Family for anti-CSTB antibody	Antibodies; Enzymes; Protease (Proteinase; Peptidase)
Applications Tested/Suitable for anti-CSTB antibody	Western Blot (WB), FLISA
Application Notes for anti-CSTB antibody	WB: 1:100-500 FLISA: 1:1,000 Applications are based on unconjugated antibody.
Related Product Information for anti-CSTB antibody	The cystatin superfamily encompasses proteins that contain multiple cystatin-like sequences. Some of the members are active cysteine protease inhibitors, while others have lost or perhaps never acquired this inhibitory activity. There are three inhibitory families in the superfamily, including the type 1 cystatins (stefins), type 2 cystatins and kininogens. This gene encodes a stefin that functions as an intracellular thiol protease inhibitor. The protein is able to form a dimer stabilized by noncovalent forces, inhibiting papain and cathepsins l, h and b. The protein is thought to play a role in protecting against the proteases leaking from lysosomes. Evidence indicates that mutations in this gene are responsible for the primary defects in patients with progressive myoclonic epilepsy (EPM1).

Product References and Citations for anti-CSTB antibody	Bailey, S.D., et al. Diabetes Care 33(10):2250-2253(2010). Ceru, S., et al. J. Biol. Chem. 285(13):10078-10086(2010). Skerget, K., et al. J. Biol. Chem. 285(5):3201-3210(2010). Ceru, S., et al. Biol. Cell 102(6):319-334(2010). Talmud, P.J., et al. Am. J. Hum. Genet. 85(5):628-642(2009).
Other Notes	Small volumes of anti-CSTB antibody vial(s) may occasionally become entrapped in the seal of the product vial during shipment and storage. If necessary, briefly centrifuge the vial on a tabletop centrifuge to dislodge any liquid in the container`s cap. Certain products may require to ship with dry ice and additional dry ice fee may apply.
Searchable Terms for CSTB purchase	MBS6289520 is an antibody targeting CSTB. Also known as CSTB, ID (CSTB, CST6, STFB, Cystatin-B, CPI-B, Liver thiol proteinase inhibitor, Stefin-B) (FITC). Reactive in Human. Host: Rabbit.



Western Blot (WB) of anti-CSTB antibody
CSTB Antibody (Center) western blot analysis in SK-BR-3 cell line lysates (35ug/lane).This demonstrates the CSTB antibody detected the CSTB protein (arrow).

ADDITIONAL DETAILS

NCBI/Uniprot data below describe general gene information for CSTB. It may not necessarily be applicable to this product.

NCBI GI #	4503117
NCBI GeneID	1476
NCBI Accession #	NP_000091.1
UniProt Primary Accession #	P04080
UniProt Related Accession #	P04080
GenBank Nucleotide #	NM_000100.3
NCBI Official Full Name	cystatin-B
NCBI Official Synonym Full Names	cystatin B (stefin B)
NCBI Official Symbol	CSTB
NCBI Official Synonym Symbols	PME; ULD; CST6; EPM1; STFB; EPM1A
UniProt Protein Name	Cystatin-B
UniProt Synonym Protein Names	CPI-B; Liver thiol proteinase inhibitor; Stefin-B
Protein Family	Cystatin
UniProt Gene Name	CSTB
UniProt Synonym Gene Names	CST6; STFB
UniProt Entry Name	CYTB_HUMAN
NCBI Protein Information	cystatin-B; CPI-B; liver thiol proteinase inhibitor

UniProt Comments for CSTB	CSTB: This is an intracellular thiol proteinase inhibitor. Tightly binding reversible inhibitor of cathepsins L, H and B. Defects in CSTB are the cause of progressive myoclonic epilepsy type 1 (EPM1). EPM1 is an autosomal recessive disorder characterized by severe, stimulus-sensitive myoclonus and tonic-clonic seizures. The onset, occurring between 6 and 13 years of age, is characterized by convulsions. Myoclonus begins 1 to 5 years later. The twitchings occur predominantly in the proximal muscles of the extremities and are bilaterally symmetrical, although asynchronous. At first small, they become late in the clinical course so violent that the victim is thrown to the floor. Mental deterioration and eventually dementia develop. Belongs to the cystatin family. Protein type: Inhibitor Chromosomal Location of Human Ortholog: 21q22.3 Cellular Component: extracellular space; cytoplasm; nucleolus Molecular Function: protease binding; endopeptidase inhibitor activity; cysteine protease inhibitor activity Biological Process: negative regulation of proteolysis; regulation of apoptosis; adult locomotory behavior; negative regulation of peptidase activity Disease: Myoclonic Epilepsy Of Unverricht And Lundborg
Research Articles on CSTB	**1.** Skull thickening and an increased prevalence of abnormal findings in skeletal radiographs of patients with EPM1 suggest that this condition is connected to defective cystatin B function.
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