

Immunotag™ Cystatin B Polyclonal Antibody

Antibody Specification	
Catalog No.	ITT6009
Product Description	Immunotag™ Cystatin B Polyclonal Antibody
Size	50 µg, 100 µg
Conjugation	HRP, Biotin, FITC, Alexa Fluor® 350, Alexa Fluor® 405, Alexa Fluor® 488, Alexa Fluor® 555, Alexa Fluor® 594, Alexa Fluor® 647
IMPORTANT NOTE	This product is custom manufactured with a lead time of 3-4 weeks. Once in production, this item cannot be cancelled from an order and is not eligible for return.
Target Protein	CySTATin B
Clonality	Polyclonal
Storage/Stability	-20°C/1 year
Application	IHC-p,ELISA
Recommended Dilution	IHC-p 1:50-200, ELISA 1:10000-20000
Concentration	1 mg/ml
Reactive Species	Human
Host Species	Rabbit
Immunogen	Synthetic peptide from human protein at AA range: 20-60
Specificity	The antibody detects endogenous Cystatin B
Purification	The antibody was affinity-purified from rabbit antiserum by affinity-chromatography using epitope-specific immunogen
Form	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Gene Name	CSTB CST6 STFB
Accession No.	P04080 Q62426
Alternate Names	Cystatin-B (CPI-B) (Liver thiol proteinase inhibitor) (Stefin-B)

Antibody Specification

Description	cystatin B(CSTB) Homo sapiens 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). One type of mutation responsible for EPM1 is the expansion in the promoter region of this gene of a CCCGCCCCGCG rep
Protein Expression	Cerebellum,Placenta,
Subcellular Localization	extracellular space,nucleolus,cytoplasm,extracellular exosome,
Protein Function	disease:Defects in CSTB are the cause of progressive myoclonic epilepsy type 1 (EPM1) [MIM:254800]. 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.,function:This is an intracellular thiol proteinase inhibitor. Tightly binding reversible inhibitor of cathepsins L, H and B.,similarity:Belongs to the cystatin family.,subunit:Able to form dimers stabilized by noncovalent forces.,
Usage	For Research Use Only! Not for diagnostic or therapeutic procedures.