



Cystatin B Monoclonal Antibody

Catalog No	BYmab-03804
Isotype	IgG
Reactivity	Human;Rat;Mouse;
Applications	WB
Gene Name	CSTB
Protein Name	Cystatin-B
Immunogen	The antiserum was produced against synthesized peptide derived from human Stefin B. AA range:49-98
Specificity	Cystatin B Monoclonal Antibody detects endogenous levels of Cystatin B protein.
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source	Monoclonal, Mouse,IgG
Purification	The antibody was affinity-purified from mouse antiserum by affinity-chromatography using epitope-specific immunogen.
Dilution	WB 1:500-2000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	CSTB; CST6; STFB; Cystatin-B; CPI-B; Liver thiol proteinase inhibitor; Stefin-B
Observed Band	11kD
Cell Pathway	Cytoplasm . Nucleus .
Tissue Specificity	Cerebellum,Placenta,
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.,

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Background

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 CCGGCCCGCG rep

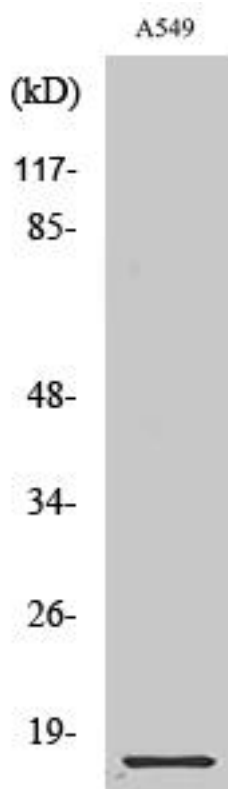
matters needing attention

Avoid repeated freezing and thawing!

Usage suggestions

This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

Products Images



Western Blot analysis of various cells using Cystatin B Monoclonal Antibody