Product Name: Cystatin B Rabbit Polyclonal Antibody
Catalog #: APRab09694

Summary

Production Name	Cystatin B Rabbit Polyclonal Antibody
Description	Rabbit Polyclonal Antibody
Host	Rabbit
Application	IHC,ELISA
Reactivity	Human,Rat,Mouse

Performance

Conjugation	Unconjugated
Modification	Unmodified
Isotype	IgG
Clonality	Polyclonal
Form	Liquid
Storage	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
Buffer	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Purification	Affinity purification

Immunogen

Gene Name	CSTB CST6 STFB
Alternative Names	Cystatin-B (CPI-B;Liver thiol proteinase inhibitor;Stefin-B)
Gene ID	1476.0
SwissProt ID	P04080.Synthetic peptide from human protein at AA range: 20-60

Application

Dilution Ratio	IHC-p 1:50-200, ELISA 1:10000-20000.
Molecular Weight	

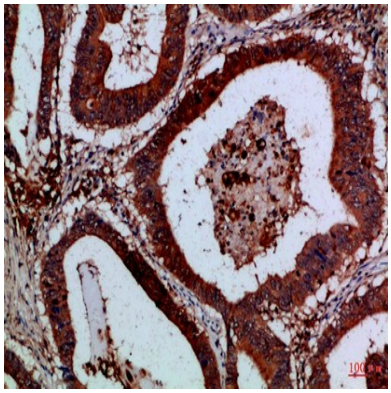
Background

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 CCCCGCCCCGCG repdisease: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.,

Research Area

Image Data



Immunohistochemical analysis of paraffin-embedded Human-colon-cancer, antibody was diluted at 1:100

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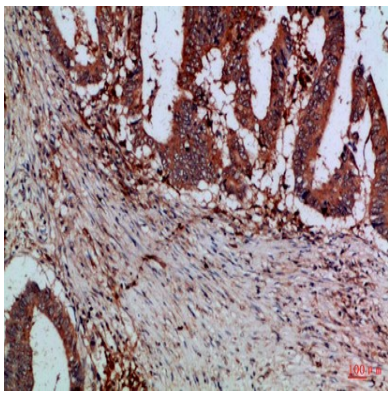

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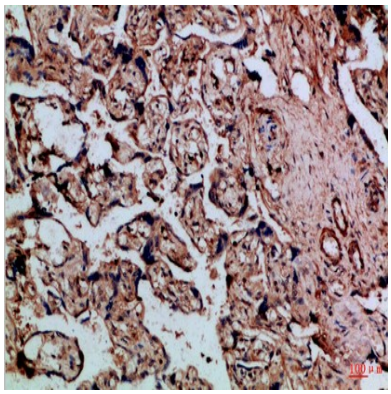

Immunohistochemical analysis of paraffin-embedded Human-placenta, antibody was diluted at 1:100

Note

For research use only.