产品名称: Cystatin B Rabbit Polyclonal Antibody

产品货号: APRab09694



产品概述 (Summary)

产品名称 (Production Name) Cystatin B Rabbit Polyclonal Antibody

描述 (Description) Rabbit polyclonal Antibody

宿主 (Host) Rabbit

应用 (Application)IHC,ICC/IF,ELISA种属反应性 (Reactivity)Human,Rat,Mouse

产品性能 (Performance)

偶联物 (Conjugation) Unconjugated 修饰 (Modification) Unmodified

同种型 (Isotype) IgG

克隆 (Clonality) Polyclonal 形式 (Form) Liquid

Store at 4°C short term. Aliquot and store at -20°C long term. Avoid 存放说明 (Storage)

freeze/thaw cycles.

Liquid in PBS containing 50% glycerol, 0.5% protective protein and 0.02% 储存溶液 (Buffer)

New type preservative N.

纯化方式 (Purification) Affinity purification

免疫原信息 (Immunogen)

基因名 (Gene Name) CSTB CST6 STFB

别名 (Alternative Names) Cystatin-B (CPI-B;Liver thiol proteinase inhibitor;Stefin-B)

基因 ID (Gene ID) 1476.0

蛋白 ID (SwissProt ID) P04080.Synthetic peptide from human protein at AA range: 20-60

产品应用(Application)

稀释比 (Dilution Ratio) IHC 1:50-1:200,ICC/IF 1:50-1:200,ELISA 1:10000-1:20000

蛋白分子量 (Molecular Weight)

研究背景 (Background)

The cystatin superfamily encompasses proteins that contain multiple cystatin-like sequences. Some of the members are

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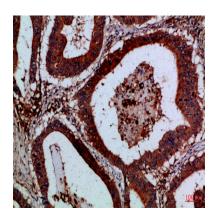


active cysteine protease inhibitors, while others have lost or perhaps never acquired this inhibitory activity. There are three inhibitory families in the superfamily, including the type 1 cystatins (stefins), type 2 cystatins and kininogens. This gene encodes a stefin that functions as an intracellular thiol protease inhibitor. The protein is able to form a dimer stabilized by noncovalent forces, inhibiting papain and cathepsins I, 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.,

研究领域 (Research Area)

Cell Biology

图片 (Image Data)



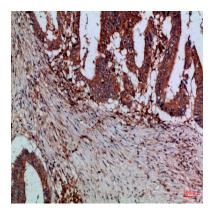
Immunohistochemical analysis of paraffin-embedded Human-colon-cancer, antibody was diluted at 1:100

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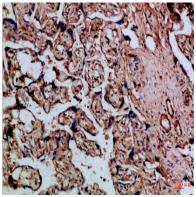
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Immunohistochemical analysis of paraffin-embedded Human-colon-cancer, antibody was diluted at 1:100



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

注意事项 (Note)

For research use only .

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