神经细胞蜡样质脂褐质沉积病蛋白 CLN5 抗体

产品货号： mlR11714

英文名称： CLN5

中文名称： 神经细胞蜡样质脂褐质沉积病蛋白 CLN5 抗体

别　　名： Ceroid lipofuscinosis neuronal 5; Ceroid-lipofuscinosis neuronal protein 5; CLN5; CLN5_HUMAN; NCL; Protein CLN5.

研究领域： 细胞生物　神经生物学

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Rat, Pig, Cow, Horse, Rabbit, Sheep,

产品应用： WB=1:500-2000 ELISA=1:500-1000
not yet tested in other applications.
optimal dilutions/concentrations should be determined by the end user.

分 子 量： 37kDa

细胞定位： 细胞浆

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from human CLN5:61-120/358

亚　　型： IgG

纯化方法： affinity purified by Protein A

储 存 液： 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件： Store at -20 ° C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20° C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 ° C.

PubMed : PubMed

产品介绍： Neuronal ceroid-lipofuscinose (NCL), also designated Batten disease, comprises a group of recessively inherited, progressive neurodegenerative diseases found in children. NCL is characterized by atrophy of the brain and an accumulation of lysosome derived fluorescent bodies found in many cells, especially neurons. Symptoms of NCL include a failure of psychomotor development, seizures, impaired vision and premature death. The eight genes/proteins associated with NCL are designated CLN1-CLN8. Mutations in six of these genes results in a distinct type of NCL-disease; the six genes/proteins are CLN1 (encoding PPT1, a protein thiolesterase), CLN2 (encodeing the serine protease TPP1), CLN3, CLN5, CLN6 and CLN8. A single base duplication mutation in dog and cow CLN5 has been shown to cause NCL.

Subcellular Location:

Lysosome.

Tissue Specificity:

Ubiquitous.

Post-translational modifications:

Glycosylated.

DISEASE:

Defects in CLN5 are the cause of neuronal ceroid lipofuscinosis type 5 (CLN5) [MIM:256731]; also known as Finnish variant late-infantile neuronal ceroid lipofuscinosis (vLINCL). A form of neuronal ceroid lipofuscinosis. Neuronal ceroid lipofuscinoses are progressive neurodegenerative, lysosomal storage diseases characterized by intracellular accumulation of autofluorescent liposomal material, and clinically by seizures, dementia, visual loss, and/or cerebral atrophy. The lipopigment patterns observed most often in neuronal ceroid lipofuscinosis type 5 comprise mixed combinations of granular, curvilinear, and fingerprint profiles.

Similarity:

Belongs to the CLN5 family.

SWISS:

O75503

Gene ID:

1203

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

产品图片

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