

产品名称: FKRP Rabbit Polyclonal Antibody

产品货号: APRab11015

产品概述 (Summary)

描述(Description)	Rabbit polyclonal Antibody
宿主(Host)	Rabbit
应用(Application)	WB,ICC/IF,ELISA
种属反应(Reactivity)	Human,Mouse,Rat
偶联物(Conjugation)	Unconjugated
修饰(Modification)	Unmodified
同种型(Isotype)	IgG
克隆(Clonality)	Polyclonal
剂型(Form)	Liquid
保存条件(Storage)	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
储存溶液(Buffer)	Liquid in PBS containing 50% glycerol, 0.5% protective protein and 0.02% New type preservative N.
纯化 (Purification)	Affinity purification

产品应用(Application)

稀释比(Dilution Ratio)	WB 1:500-1:2000,ICC/IF 1:200-1:1000,ELISA 1:10000-1:20000
分子量(Molecular Weight)	50kDa

抗原信息(Antigen Information)

基因名(Gene Name)	FKRP
别名(Alternative Names)	FKRP; Fukutin-related protein
基因 ID(Gene ID)	79147.0
SwissProt ID	Q9H9S5
免疫原(Immunogen)	The antiserum was produced against synthesized peptide derived from human FKRP. AA range:1-50

研究背景 (Background)

This gene encodes a protein which is targeted to the medial Golgi apparatus and is necessary for posttranslational modification of dystroglycan. Mutations in this gene have been associated with congenital muscular dystrophy, mental retardation, and

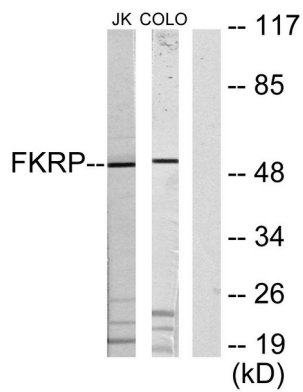
cerebellar cysts. Several alternatively spliced transcript variants of this gene have been described, but the full-length nature of some of these variants has not been determined. [provided by RefSeq, Oct 2008],disease:Defects in FKRP are the cause of congenital muscular dystrophy type 1C (MDC1C) [MIM:606612]. Congenital muscular dystrophies (CMD) are a heterogeneous group of autosomal recessive disorders characterized by hypotonia, muscle weakness, and joint contractures that present at birth or during the first 6 months of life and have dystrophic changes on skeletal muscle biopsy. Mental retardation with or without structural CNS changes may accompany some forms. MDC1C is a form of CMD with onset in the first weeks of life and a severe phenotype with inability to walk, muscle hypertrophy, marked elevation of serum creatine kinase, a secondary deficiency of laminin alpha2, and a marked reduction in alpha-dystroglycan expression. Only a subset of MDC1C patients have brain involvements.,disease:Defects in FKRP are the cause of limb-girdle muscular dystrophy type 2I (LGMD2I) [MIM:607155]. LGMD2I is an autosomal recessive disorder with age of onset ranging from childhood to adult life, and variable severity. Clinical features include proximal muscle weakness, waddling gait, calf hypertrophy, cardiomyopathy and respiratory insufficiency. A reduction of alpha-dystroglycan and laminin alpha-2 expression can be observed on skeletal muscle biopsy from LGMD2I patients.,disease:Defects in FKRP may be a cause of muscle-eye-brain disease (MEB) [MIM:253280]. MEB is an autosomal recessive disorder characterized by congenital muscular dystrophy, ocular abnormalities, cobblestone lissencephaly and cerebellar hypoplasia. MEB patients present severe congenital myopia, congenital glaucoma, pallor of the optic disks, retinal hypoplasia, mental retardation, hydrocephalus, abnormal electroencephalograms, generalized muscle weakness and myoclonic jerks.,disease:Defects in FKRP may be a cause of Walker-Warburg syndrome (WWS) [MIM:236670]; also known as hydrocephalus-agyria-retinal dysplasia or HARD syndrome. WWS is an autosomal recessive disorder characterized by cobblestone lissencephaly, hydrocephalus, agyria, retinal dysplasia, with or without encephalocele. It is often associated with congenital muscular dystrophy and usually lethal within the first few months of life.,function:Could be a transferase involved in the modification of glycan moieties of alpha-dystroglycan (DAG1),online information:GlycoGene database,similarity:Belongs to the licD transferase family.,tissue specificity:Expressed predominantly in skeletal muscle, placenta, and heart and relatively weakly in brain, lung, liver kidney and pancreas.,

研究领域 (Research Area)

注意事项 (Note)

For research use only.

图片 (Image Data)



Western blot analysis of lysates from Jurkat and COLO205 cells, using FKRP Antibody. The lane on the right is blocked with the synthesized peptide.

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产品类别	产品货号	产品名称
WB 解决方案	RA10020	2h 极速 WB 即用型全流程试剂盒
	RA10021	4h 快速 WB 即用型全流程试剂盒
	RA10042	5h 畅享版 WB 全流程试剂盒
	RA10037	校准级彩色预染蛋白 Marker (8-180kDa)
	RA10038	校准级彩色预染蛋白 Marker (10-250kDa)
	RA10039	校准级高分子彩色预染蛋白 Marker (25-400kDa)
TSA 多重荧光染色试剂盒	RA10008	TSA 双标三色多重荧光染色试剂盒 (mIHC)
	RA10009	TSA 三标四色多重荧光染色试剂盒 (mIHC)
	RA10010	TSA 四标五色多重荧光染色试剂盒 (mIHC)
	RA10011	TSA 五标六色多重荧光染色试剂盒 (mIHC)
	RA10012	TSA 六标七色多重荧光染色试剂盒 (mIHC)
IHC 检测试剂盒	RA10006	HRP Anti-Mouse/Rabbit IHC Detection System
	RA10007	Polymer-HRP Anti-Mouse/Rabbit IHC Detection System
抗体标记试剂盒	RE80004p	辣根过氧化物酶(HRP)抗体标记试剂盒
	RE80002q	Sulfo-NHS-生物素标记试剂盒
	RE80007p	Cy3 荧光素标记试剂盒
	RE80011p	Fluor488 荧光素标记试剂盒
	RE80017p	Fluor750 荧光素标记试剂盒
	RE80005p	藻红蛋白(R-PE) 抗体快速标记试剂盒
	RE80040	PE-Cy7 串联染料抗体快速标记试剂盒
稳转细胞系构建服务 (免费赠送全膜 WB 验证)	TS-0001	过表达稳转细胞系构建
	TS-0002	敲低稳转细胞系构建
	TS-0003	敲除细胞系构建

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