

产品名称: ATPAF2 Rabbit Polyclonal Antibody
产品货号: APRab07346



产品概述 (Summary)

产品名称 (Production Name)	ATPAF2 Rabbit Polyclonal Antibody
描述 (Description)	Rabbit polyclonal Antibody
宿主 (Host)	Rabbit
应用 (Application)	WB,ELISA
种属反应性 (Reactivity)	Human,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
存放说明 (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.
纯化方式 (Purification)	Affinity purification

免疫原信息 (Immunogen)

基因名 (Gene Name)	ATPAF2
别名 (Alternative Names)	ATPAF2; ATP12; LP3663; ATP synthase mitochondrial F1 complex assembly factor 2; ATP12 homolog
基因 ID (Gene ID)	91647.0
蛋白 ID (SwissProt ID)	Q8N5M1.The antiserum was produced against synthesized peptide derived from human ATPAF2. AA range:21-70

产品应用 (Application)

稀释比 (Dilution Ratio)	WB 1:500-1:2000,ELISA 1:20000-1:40000
蛋白分子量 (Molecular Weight)	35kDa

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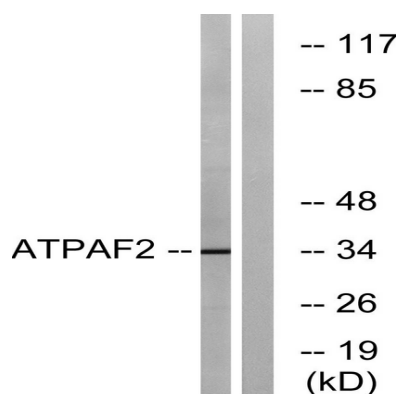


研究背景 (Background)

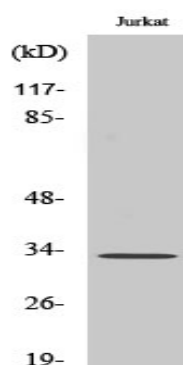
ATP synthase mitochondrial F1 complex assembly factor 2(ATPAF2) Homo sapiens This gene encodes an assembly factor for the F(1) component of the mitochondrial ATP synthase. This protein binds specifically to the F1 alpha subunit and is thought to prevent this subunit from forming nonproductive homooligomers during enzyme assembly. This gene is located within the Smith-Magenis syndrome region on chromosome 17. An alternatively spliced transcript variant has been described, but its biological validity has not been determined. [provided by RefSeq, Jul 2008],disease:Defects in ATPAF2 are the cause of complex V mitochondrial respiratory chain ATPAF2 subunit deficiency (ATPAF2 deficiency) [MIM:604273]; also called ATP synthase deficiency or ATPase deficiency. ATPAF2 deficiency seems to be an early presenting disease in which lactic acidosis, dysmorphic features, and methyl glutaconic aciduria can be major clues in the diagnosis. Dysmorphic features include a large mouth, prominent nasal bridge, micrognathia, rocker-bottom feet and flexion contractures of the limbs associated with camptodactyly. Patients are hypertonic and have an enlarged liver, hypoplastic kidneys and elevated lactate levels in urine, plasma and cerebro spinal fluid (CSF).,function:May play a role in the assembly of the F1 component of the mitochondrial ATP synthase (ATPase).,similarity:Belongs to the ATP12 family.,subunit:Interacts with ATP5A1.,tissue specificity:Widely expressed.,

研究领域 (Research Area)

图片 (Image Data)



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Western Blot analysis of various cells using ATPAF2 Polyclonal Antibody

注意事项 (Note)

For research use only .