



# ATPAF2 Monoclonal Antibody

Catalog No	BYmab-16394
Isotype	IgG
Reactivity	Human;Mouse
Applications	WB
Gene Name	ATPAF2
Protein Name	ATP synthase mitochondrial F1 complex assembly factor 2
Immunogen	The antiserum was produced against synthesized peptide derived from human ATPAF2. AA range:21-70
Specificity	ATPAF2 Monoclonal Antibody detects endogenous levels of ATPAF2 protein.
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source	Monoclonal, Mouse,IgG
Purification	The antibody was affinity-purified from mouse antiserum by affinity-chromatography using epitope-specific immunogen.
Dilution	WB 1:500-2000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	ATPAF2; ATP12; LP3663; ATP synthase mitochondrial F1 complex assembly factor 2; ATP12 homolog
Observed Band	35kD
Cell Pathway	Mitochondrion .
Tissue Specificity	Widely expressed.
Function	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

Nanjing BYabscience technology Co.,Ltd



mitochondrial ATP synthase (ATPase).,similarity:Belongs to the ATP12 family.,subunit:Interacts with ATP5A1.,tissue specificity:Widely expr

#### 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],

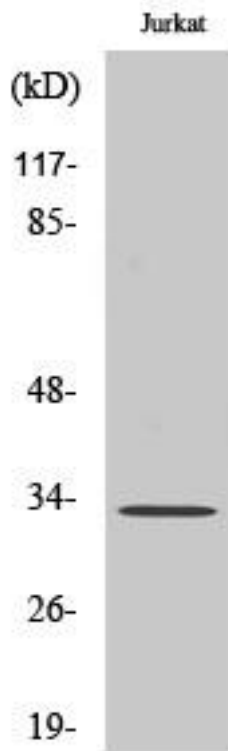
#### matters needing attention

Avoid repeated freezing and thawing!

#### Usage suggestions

This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

### Products Images



Western Blot analysis of various cells using ATPAF2 Monoclonal Antibody