Product Name: ATPAF2 Rabbit Polyclonal Antibody
Catalog #: APRab07346

Summary

Production Name	ATPAF2 Rabbit Polyclonal Antibody
Description	Rabbit Polyclonal Antibody
Host	Rabbit
Application	WB,ELISA
Reactivity	Human,Mouse

Performance

Conjugation	Unconjugated
Modification	Unmodified
Isotype	IgG
Clonality	Polyclonal
Form	Liquid
Storage	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
Buffer	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Purification	Affinity purification

Immunogen

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

Application

Dilution Ratio	WB 1:500 - 1:2000. ELISA: 1:40000.
Molecular Weight	35kD

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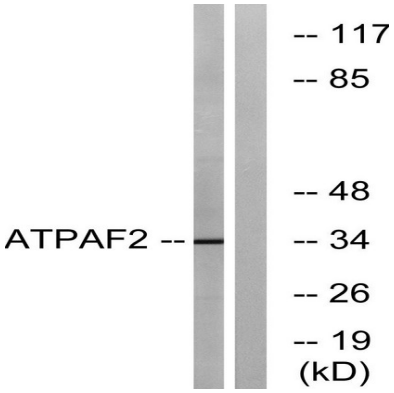
Background

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.,

Research Area

Image Data



Western blot analysis of lysates from Jurkat cells, using ATPAF2 Antibody. The lane on the right is blocked with the synthesized peptide.

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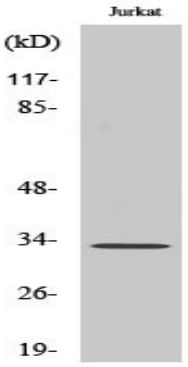

Western Blot analysis of various cells using ATPAF2 Polyclonal Antibody

Note

For research use only.