



AL4A1 rabbit pAb

Catalog No	YP-Ab-07989
Isotype	IgG
Reactivity	Human; Mouse;Rat
Applications	WB
Gene Name	ALDH4A1 ALDH4 P5CDH
Protein Name	AL4A1
Immunogen	Synthesized peptide derived from human AL4A1 AA range: 39-89
Specificity	This antibody detects endogenous levels of AL4A1 at Human/Mouse/Rat
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.103% sodium azide.
Source	Polyclonal, Rabbit,IgG
Purification	The antibody was affinity-purified from rabbit serum by affinity-chromatography using specific immunogen.
Dilution	WB 1:500-2000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	Delta-1-pyrroline-5-carboxylate dehydrogenase, mitochondrial (P5C dehydrogenase) (EC 1.5.1.12) (Aldehyde dehydrogenase family 4 member A1)
Observed Band	60kD
Cell Pathway	Mitochondrion matrix.
Tissue Specificity	Highest expression is found in liver followed by skeletal muscle, kidney, heart, brain, placenta, lung and pancreas.
Function	catalytic activity:(S)-1-pyrroline-5-carboxylate + NAD(P)(+) + 2 H(2)O = L-glutamate + NAD(P)H.;disease:Defects in ALDH4A1 are the cause of hyperprolinemia type II (HPH) [MIM:239510]. HPH is characterized by the accumulation of delta-1-pyrroline-5-carboxylate (P5C) and proline. The disorder may be causally related to neurologic manifestations, including seizures and mental retardation.;function:Irreversible conversion of delta-1-pyrroline-5-carboxylate (P5C), derived either from proline or ornithine, to glutamate. This is a necessary step in the pathway interconnecting the urea and tricarboxylic acid cycles. The preferred substrate is glutamic gamma-semialdehyde, other substrates include succinic, glutaric and adipic semialdehydes.;pathway:Amino-acid degradation; L-proline degradation into L-glutamate; L-glutamate from L-proline: step 2/2.;similarity:Belongs to the aldehyde dehydrogenase



Background

This protein belongs to the aldehyde dehydrogenase family of proteins. This enzyme is a mitochondrial matrix NAD-dependent dehydrogenase which catalyzes the second step of the proline degradation pathway, converting pyrroline-5-carboxylate to glutamate. Deficiency of this enzyme is associated with type II hyperprolinemia, an autosomal recessive disorder characterized by accumulation of delta-1-pyrroline-5-carboxylate (P5C) and proline. Alternatively spliced transcript variants encoding different isoforms have been identified for this gene. [provided by RefSeq, Jun 2009],

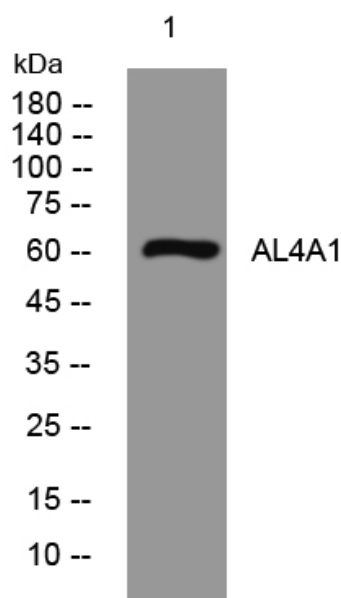
matters needing attention

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 lysates from A431 cells, primary antibody was diluted at 1:1000, 4° over night