

# Recombinant Human ALDH4A1 Protein

Catalog Number:PKSH030775



**Note:** Centrifuge before opening to ensure complete recovery of vial contents.

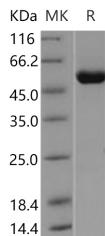
## Description

<b>Synonyms</b>	ALDH4;P5CD;P5CDh
<b>Species</b>	Human
<b>Expression Host</b>	Baculovirus-Insect Cells
<b>Sequence</b>	Lys 25-Gln 563
<b>Accession</b>	AAH07581.1
<b>Calculated Molecular Weight</b>	59.2 kDa
<b>Observed molecular weight</b>	54 kDa
<b>Tag</b>	None

## Properties

<b>Purity</b>	> 95 % as determined by reducing SDS-PAGE.
<b>Endotoxin</b>	< 1.0 EU per $\mu$ g of the protein as determined by the LAL method.
<b>Storage</b>	Generally, lyophilized proteins are stable for up to 12 months when stored at -20 to -80°C. Reconstituted protein solution can be stored at 4-8°C for 2-7 days. Aliquots of reconstituted samples are stable at < -20°C for 3 months.
<b>Shipping</b>	This product is provided as lyophilized powder which is shipped with ice packs.
<b>Formulation</b>	Lyophilized from sterile 20mM Tris, 500mM NaCl, 10% glycerol, pH 8.5 Normally 5 % - 8 % trehalose, mannitol and 0.01% Tween80 are added as protectants before lyophilization. Please refer to the specific buffer information in the printed manual.
<b>Reconstitution</b>	Please refer to the printed manual for detailed information.

## Data



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## Background

ALDH4A1 is a member of the aldehyde dehydrogenase family. Aldehyde dehydrogenase enzymes function in the metabolism of many molecules including certain fats (cholesterol and other fatty acids) and protein building blocks (amino acids). Additional aldehyde dehydrogenase enzymes detoxify external substances, such as alcohol and pollutants, and internal substances, such as toxins that are formed within cells. ALDH4A1 is expressed abundantly in liver followed by skeletal muscle, kidney, heart, brain, placenta, lung and pancreas. It is a mitochondrial matrix NAD-dependent dehydrogenase which catalyzes the second step of the proline degradation pathway, converting pyrroline-5-carboxylate to glutamate. Defects in ALDH4A1 are the cause of hyperprolinemia type 2 (HP-2). HP-2 is characterized by the accumulation of delta-1-pyrroline-5-carboxylate (P5C) and proline. The disorder may be causally related to neurologic

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manifestations, including seizures and mental retardation.

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