

**Recombinant Human ALDH4A1 Protein**

**Cat.NO.: TP06954**

3th Edition

**Synonyms:**ALDH4;P5CD;P5CDh

**Description:**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 manifestations, including seizures and mental retardation.

**Form:**PBS

**Molecular Weight:**59.2 kDa

**Sequences:**Lys 25-Gln 563

**Purity:**> 95% by HPLC

**Concentration:**

**Endotoxin Level:**<1.0 EU per 1 ug of protein (determined by LAL method)

**Storage:**Can be stored at +4°C short term (1-2 weeks). For long term storage, aliquot and store at -20°C or -70°C. Avoid repeated freezing and thawing cycles.